REPORT

Alberta Congenital Anomalies Surveillance System Seventh Report 1980-2005

Alberta

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ALBERTA CONGENITAL ANOMALIES SURVEILLANCE SYSTEM

SEVENTH REPORT

1980 – 2005

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1 EXECUTIVE SUMMARY

1. This is the seventh in a series of reports detailing the prevalence of congenital anomalies in Alberta dealing particularly with the years 2000 to 2004 inclusive. Aggregate data is also included from 1980 onwards.

2. The International Classification of Diseases – 10th Edition (ICD-10) classification system has been adopted by Alberta and the report uses the Royal College of Paediatrics and Child Health adaptation of ICD-10. The anomalies outlined in the National Birth Defects Prevention Network's Guidelines for Conducting Birth Defects Surveillance (http://www.nbdpn.org/index.html) are reported in this report. However, all items from the ICD-10 "Q" codes as well as other sections such as disorders of metabolism are monitored by ACASS. Data on such disorders will be provided to interested parties upon request.

3. The overall frequency of most congenital anomalies remains relatively stable with the exception of neural tube defects which continue to decline. The decline is most obvious with anencephaly although there is a significant downward trend with spina bifida as well. Although some of the decline might be attributable to termination of pregnancy, folic acid fortification may also be playing a role.

4. The percentage of births to women 35 years of age and over has stabilized over the past 3 years. About 14.5% of women 35 years of age and over gave birth in the period 2000-2005.

5. Limb reductions do seem to be increasing overall and ACASS will conduct a review of these anomalies in the coming months. Limb reductions include very heterogeneous categories ranging from the absence of part of a finger to a missing arm or leg.

6. Abdominal wall defects overall are not increasing significantly. However, when one examines the defects included in the category, an increase in the rate of gastroschisis is noted. On the other hand omphalocele rates have been very stable over the years. The occurrence of gastroschisis is more frequent in younger mothers in Alberta which is consistent with observations from other jurisdictions.

7. Consistent with the literature, women giving birth at age 35 years or over have an increased birth prevalence rate of congenital anomalies, particularly for Down syndrome, ventricular septal defects, nervous system defects, cleft palate, renal dysgenesis, congenital hip dislocation, and abdominal wall defects. Women giving birth before age 20 years have an increased rate of birth prevalence for ventricular septal defects, cleft lip +/- cleft palate, and gastroschisis.

8. In Alberta, about 19% of infants with congenital anomalies are low birth weight. Most anomaly groups show increasing birth prevalence with decreasing birth weight, except congenital hip dislocation, ventricular septal defects, diaphragmatic hernia, gastroschisis, and hypospadias and epispadias for which the birth prevalence rate in babies weighing less than 1000 grams at birth is not higher than babies weighing 1000-1499 grams.

9. The birth prevalence of congenital anomalies varies by health region, which is partly due to regional variations in case ascertainment and reporting. Differences in demographics and other

factors may also account for some of the observed regional variations. Continued monitoring of the regional patterns is required.

10. The death rate of all congenital anomalies in Alberta has decreased from 24.3 per 10000 live births in 1983-85 to 17.4 in 2001-2003. The decrease is particularly significant for heart defects, neural tube defects (NTDs), and respiratory anomalies.

11. Because differences were noted between ACASS and Canadian Congenital Anomalies Surveillance System (CCASS) rates for anorectal atresia/stenosis, a detailed review was carried out. Results indicate that a substantial number of cases belong in the multiple congenital anomaly category such as VATER/VACTERL. A more in depth account is found in this report and a manuscript has been accepted by the Journal of Pediatric Surgery (in press).

12. ACASS continues to be a member of the Canadian Congenital Anomalies Surveillance Network (CCASN), a Public Health Agency of Canada initiative, and members of ACASS play a significant role in Network committees. The Network has been formed to support the development and maintenance of high quality population-based surveillance systems for congenital anomalies.

13. ACASS has participated in a national study on the impact of folic acid fortification on the rates of neural tube defects for the years 1993-2002. The study was funded by Canadian Institutes of Health Research (CIHR) and includes data from Newfoundland, Nova Scotia, Prince Edward Island, Quebec, Manitoba, Alberta and British Columbia. A manuscript has been submitted for publication.

14. ACASS continues its affiliation with the International Clearinghouse for Birth Defects Surveillance and Research (ICBDSR) and has participated in or is participating in a number of studies with these organizations such as the epidemiology of craniofacial defects, epidemiology of very rare defects, epidemiology of abdominal wall defects, and geographical variation of common malformations.

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2. INTRODUCTION

This Report provides data on congenital anomalies ascertained in Alberta from the years 1980-2005 inclusive. The most recent published report by Alberta Health and Wellness contains statistics up to the end of 2001. This report presents updated data from the years 1980-2001 as well as the addition of the years 2002-2005. For the current release the anomalies outlined in the National Birth Defects Prevention Network's Guidelines for Conducting Birth Defects Surveillance (2004) are reported, however, data on other anomalies can be provided upon request.

2.1 History

The history of the Alberta Congenital Anomalies Surveillance System (ACASS) has been well described in previous reports. Funding from Alberta Health and Wellness, Public Health Surveillance and Environmental Health remains stable. ACASS continues to work closely with Alberta Vital Statistics and relies on them for the provision of notifications of births, deaths and stillbirths.

2.2 Purpose of a Surveillance System

Public health surveillance in general has been defined by the Centers for Disease Control and Prevention (CDC) in Atlanta, Georgia as the ongoing, systematic collection, analysis and interpretation of data (e.g. regarding agent/hazard, risk factor, exposure, health event) essential to the planning, implementation and evaluation of public health practice, closely integrated with the timely dissemination of these data to those responsible for prevention and control.

The purposes and objectives of surveillance for congenital anomalies (CAs) are to:

1) provide reliable and valid baseline data of congenital anomalies in Alberta;
2) investigate any significant temporal or geographic changes in the frequency of congenital anomalies with a view to identifying environmental, and therefore, possibly preventable causes;
3) measure trends;
4) assess the effectiveness of prevention (e.g. folic acid fortification or antenatal screening);
5) assist with health related programme planning and development through the provision of data;
6) participate in research into the aetiology and natural history of birth defects;
7) assist researchers through provision of congenital anomalies data; and
8) provide advice to health care professionals about congenital anomalies especially with respect to teaching and launching public health campaigns (e.g. folic acid campaign by Community Health in Calgary).

As well as the above, patterns or associations of malformations to determine whether they belong to an existing or new syndrome complex can be explored.

A principle feature of a surveillance system is timeliness; however data collection and analysis should not be accomplished at the expense of an accurate diagnosis. Since data are collected by the 1st birthday, plus the possibility of delay in reporting, the data of a given calendar year may not be complete until at least the December 31 of the subsequent year.

3. METHODOLOGY

3.1 Case Definitions

A **congenital anomaly** is an abnormality that is present at birth, even if not diagnosed until months or years later. Most congenital anomalies are present long before the time of birth, some in the embryonic period (up to the end of the 7th week of gestation) and others in the fetal period (8th week to term). The term "anomaly" covers all the major classes of abnormalities of development, of which there are four major categories as follows:

Malformation – a morphologic defect of an organ, part of an organ or a larger region of the body resulting from an intrinsically abnormal developmental process (e.g. spina bifida, cleft lip and palate).
Deformation – an abnormal form, shape or position of a part of the body caused by mechanical forces (e.g. extrinsic force such as intrauterine constraint causing some forms of clubfoot).
Disruption – a morphologic defect of an organ, part of an organ or a larger region of the body resulting from the extrinsic breakdown of, or an interference with, an originally normal developmental process (e.g. an infection such as rubella or a teratogen such as thalidomide).
Dysplasia – the abnormal organization of cells into tissues and its morphologic result (e.g. Marfan Syndrome, osteogenesis imperfecta).

Other definitions related to pregnancy outcomes for the purposes of this report are as follows:

Live birth – A complete expulsion or extraction from the mother, *irrespective* of the duration of the pregnancy, of a foetus in which, after expulsion or extraction, there is breathing, beating of the heart, pulsation of the umbilical cord or definite movement of voluntary muscle (Alberta Vital Statistics Annual review, 2000).
Stillbirth – A complete expulsion or extraction from the mother, after at least 20 weeks pregnancy (≥20 weeks), *or* after attaining a weight of 500 grams or more (≥500 grams) of a foetus in which, after the expulsion or extraction, there is no breathing, beating of the heart, pulsation of the umbilical cord or unmistakable movement of voluntary muscle (Alberta Vital Statistics Annual review, 2000).
Gestation – Completed weeks at delivery.
Preterm birth (aka premature) – An infant born before 37 weeks of gestation (<37 weeks).
Termination of Pregnancy (ToP) – For our purposes, any pregnancy loss before 20 weeks of gestation (<20 weeks) most of which are therapeutic terminations for congenital anomalies but could include spontaneous abortions or intrauterine foetal deaths with foetal anomalies.

3.2 Case Ascertainment

An infant can be ascertained at any time up to the first birthday. Multiple ascertainment of the same infant can occur and is encouraged, as this frequently improves the quality and reliability of the data.

As several malformations may occur in the same infant, it is advantageous to allow each to be reported so that groups of associated malformations may be studied. This, however, leads to difficulties since the final tabulations may be reported as total malformations (anomaly rates) or as the total number of malformed infants (case rates).

ACASS obtains information about infants with congenital anomalies from a variety of independent sources. Acquisition of additional reporting agencies is always a priority since the use of multiple sources of information improves not only the ease but also completeness of ascertainment as well as accuracy of the diagnostic data. **Appendix A.1** shows the process of data collection at ACASS.

ACASS screens many important Alberta Health and Wellness and Alberta Vital Statistics documents for the presence of a congenital anomaly including:

- Notice of a Live birth or a Stillbirth and Newborn Record often referred to as the Physician's Notice of Birth (PNOB)
- Medical Certificate of Stillbirth
- Medical Certificate of Death

Also, ACASS screens a notification called the Congenital Anomalies Reporting Form (CARF, **Appendix A.2**) that is completed by all acute care hospitals in the province on live births, stillbirths, admissions or hospital deaths of infants under one year of age as well as pregnancy losses involving one or more congenital anomalies. This form serves as the single most important source of case ascertainment.

Since many children with congenital anomalies are not admitted to hospital, it is very important to obtain out-patient information such as from the Calgary and Edmonton Departments of Medical Genetics.

Ascertainment at a continued high level requires each hospital record department and each health care provider to co-operate with the system by notifying us as promptly as possible. We are fortunate in having such co-operative agencies and personnel.

3.3 Quality Control Measures

When a copy of a reporting document reaches the ACASS office in Calgary, it is reviewed for content by the Research Assistant and Manager. If the information is unclear, the Manager, on behalf of the Medical Consultant, writes to the physician responsible for the case seeking clarification. A stamped, addressed envelope is included with the letter and the physician is asked to respond at the bottom of the letter thus making the mechanics of replying easy. The response from physicians has been very satisfactory (greater than 90%) and usually this is sufficient to make a decision whether to accept or reject an anomaly or case. Any questionable diagnosis that is not confirmed is not entered into the database. Some cases also not included contain diagnoses that do not belong in a congenital anomaly system or are part of a normal developmental process such as a patent ductus arteriosus or undescended testes in a premature infant. Any reports requiring a medical decision are reviewed with the Medical Consultant. Policy decisions with respect to the acceptance or rejection of a case and its coding are referred to the ACASS Advisory Committee. This body is comprised of a paediatric cardiologist, neonatologist/epidemiologist, paediatric pathologist, medical geneticist (medical consultant) with occasional input from a paediatric neurologist, paediatric nephrologist and a paediatric orthopaedic surgeon.

3.4 Anomaly Coding

Coding is done at the Calgary office using the Royal College of Paediatrics and Child Health (RCPCH) adaptation of the International Classification of Diseases, tenth edition (ICD-10). Difficult cases are referred to the Medical Consultant (Medical Geneticist). In the past, we were able to code only 6 anomalies per case but since 1997 we have been coding all eligible anomalies reported to us.

3.5 Data Linkage and Maternal Risk Factor Data

Data from ACASS are linked to data from the Vital Statistics Birth registry by the birth registration number, with over 99% success. Some maternal risk factor data, such as maternal smoking, drinking and use of street drugs during pregnancy are thus available for babies with congenital anomalies. This linkage enables in-depth data analysis and interpretation.

3.6 Confidentiality and Release of Data

Notifications of Congenital Anomalies are sent to the Public Health Surveillance and Environmental Health Branch, Alberta Health and Wellness and from there to the ACASS office in Calgary where the database is maintained. The notifications are handled by the Manager, Research Assistant, Secretary, Clerk and Medical Consultant. The data are treated in a completely confidential manner and the notifications are kept in locked files in a locked room. The database is secured by limited access and is password protected. Should further clarification about a case or anomaly become necessary, we communicate with the attending physician or the physician responsible for ongoing care. Direct contact is never made with the family. When data are requested from us, they are released in aggregate form with no personal identifiers.

3.7 Epidemiological and Statistical Measures

Unless otherwise stated, the birth defect rates presented in this report are calculated using the following formulae:

$$\text{ANOMALY (DEFECT) RATE} = \frac{\text{Number of a particular congenital anomaly among live births and stillbirths}}{\text{Total number of live births and stillbirths}} \times 1000$$

$$\text{CASE RATE} = \frac{\text{Number of individual infants (live- or stillborn) with} \geq 1 \text{ congenital anomaly}}{\text{Total number of live births and stillbirths}} \times 1000$$

Confidence intervals (95 per cent) are also included because the rate obtained is actually only a point estimate of the unknown, true population rate. The confidence interval provides information about the precision of the estimate. Thus, the confidence intervals are an estimated range of values within which there is a 95% probability that the true population rate will fall.

Linear trend analysis was performed and presented as appropriate.

3.8 Limitations of Data and Analysis

One of the major limitations of the surveillance system is that on its own, the information provided to us does not allow studies to determine aetiology. If increasing trends indicate there is a potentially serious problem, then separate investigative studies need to be done. However, it is possible to conduct linkage studies with other data sources to explore potential causes of specific birth defects.

The ACASS data are collected passively from Vital Statistics, hospitals, and other agencies but are augmented by specific inquiries to physicians and labs, etc. The completeness and accuracy of data are largely dependent on reporting. Regional variations in anomaly and case rates may be explained in part by ascertainment and reporting differences. As such, caution has to be exercised in the interpretation of regional data.

Some anomaly groups have a small number of cases thus are combined to allow stable rate estimations and meaningful comparisons. The specific methods used for each section are included in the last portion of the section.

4 PATTERNS OF SELECTED CONGENITAL ANOMALIES IN ALBERTA

4.1 Birth Prevalence – Time Trends

The following table and graphs of selected sentinel anomalies indicate the trends in congenital anomaly rates in Alberta from 1980 through 2005. Sentinel anomalies are those which the International Clearinghouse of Birth Defects Surveillance and Research (ICBDSR), of which we are a member, watches worldwide with the rationale that they are quite easily identified hence accurately reported.

Table 4.1 Chi Squared Linear Trend Analysis and p-values for Selected Anomalies 1980-2005 Inclusive† (Live Births and Stillbirths)

Anomaly	Trend Direction	Chi Squared Analysis (χ^2LT)	p-value
Neural Tube Defects	Decreasing	34.14	0.0000
Anencephaly	Decreasing	31.43	0.0000
Spina Bifida	Decreasing	15.22	0.0001
Hydrocephalus	No significant change	2.06	0.1512
Cleft Lip +/- Cleft Palate	No significant change	0.55	0.4583
Cleft Palate	Increasing	5.74	0.0166
Oesophageal Atresia/Stenosis	No significant change	3.31	0.0689
Anorectal & Large Intestine Atresia/Stenosis	Increasing	5.77	0.0163
Hypospadias and Epispadias *	No significant change	0.23	0.6315
Limb Reductions	Increasing	9.44	0.0021
Gastroschisis	Increasing	30.64	0.0000
Omphalocele	No significant change	0.21	0.6468
Down Syndrome	Increasing	56.13	0.0000
Renal Agenesis	Increasing	4.73	0.0296
Hypoplastic Left Heart Syndrome	No significant change	1.36	0.2435

*Hypospadias and Epispadias calculated for male live births only
† Data for 2005 might not be complete

4.1.1 Neural Tube Defects

Neural tube defect rates have declined in Alberta since 1980 especially anencephaly and spina bifida. Encephalocele rates have remained fairly constant. Terminations of affected pregnancies and early foetal losses might account for part of the decline but it is unlikely that these factors alone explain the total reduction of NTDs in Alberta. We have been able to follow terminations of pregnancy (ToPs) and early foetal losses since 1997. However, if we extrapolate our ToP data back through the years, there seems to be a true decline in the rates of NTDs since the 1998 introduction of folic acid fortification of flour and cereal/grain products (150 µg/100gm, **Figure 4.1.1, 4.1.2**). We have participated in the seven-province study of the impact of folic acid fortification and the prevalence of neural tube defects. Data collection is complete and the manuscript is submitted for publication. Similar observations about declining neural tube defect rates have been noted among the other participating provinces.

Figure 4.1.1 Neural Tube Defects in Total Births (Live+Still), Alberta, 1980-2005

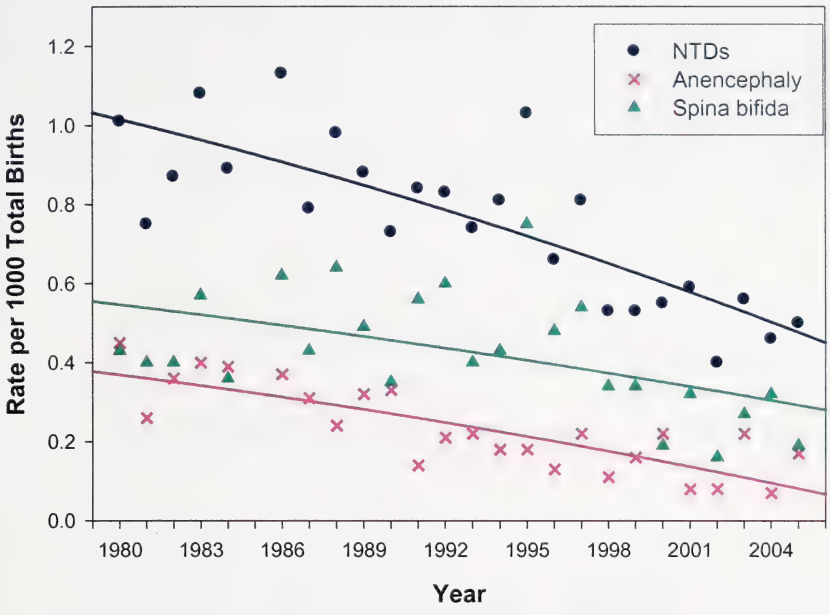
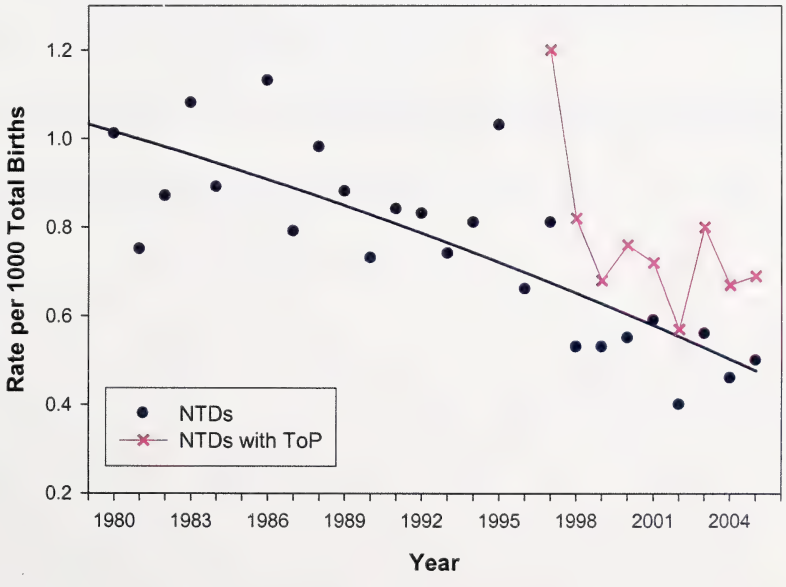


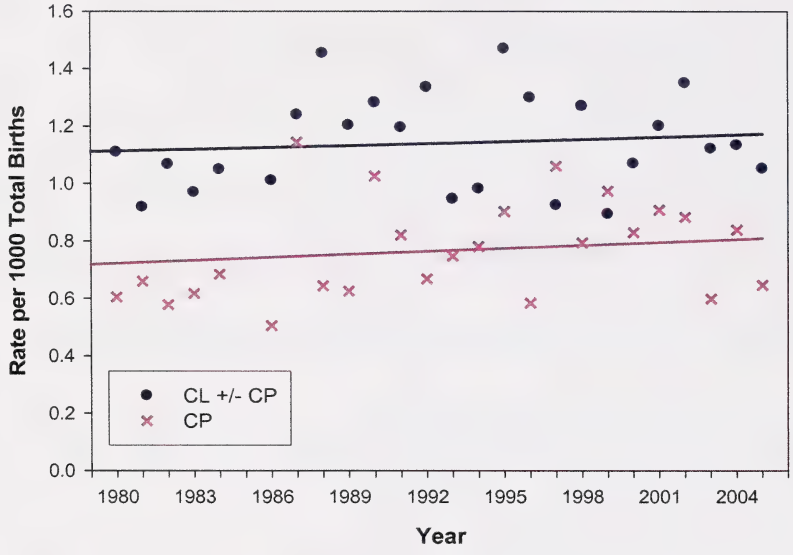
Figure 4.1.2 Neural Tube Defects including ToPs/Foetal Losses, Alberta, 1980-2005



4.1.2 Cleft Lip and Palate

The birth prevalence of cleft lip with or without cleft palate (CL ± CP) remains stable (**Figure 4.1.3, 4.1.4**). However, there does appear to be an increasing trend in the rates of cleft palate alone (CP, **Figure 4.1.5**). This could be due in part to better ascertainment. ACASS is participating in an international study of craniofacial anomalies coordinated through the International Clearinghouse of Birth Defect Surveillance and Research (ICBDSR).

Figure 4.1.3 Cleft Lip +/- Cleft Palate (CL+/-CP) and Cleft Palate (CP) alone in Total Births Alberta, 1980-2005



The rates change only minimally in some years when ToPs are added but not enough to alter the trends in either case (CL+/-CP or CP alone).

Figure 4.1.4 Cleft Lip +/- Cleft Palate including ToPs/Foetal Losses, 1980-2005

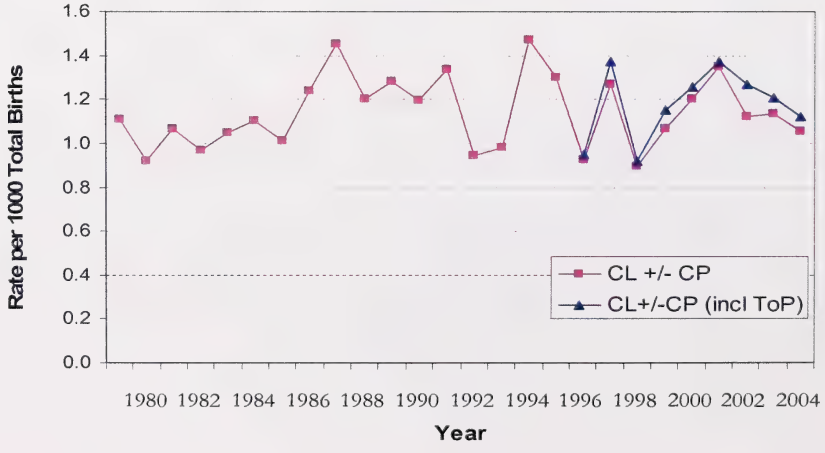
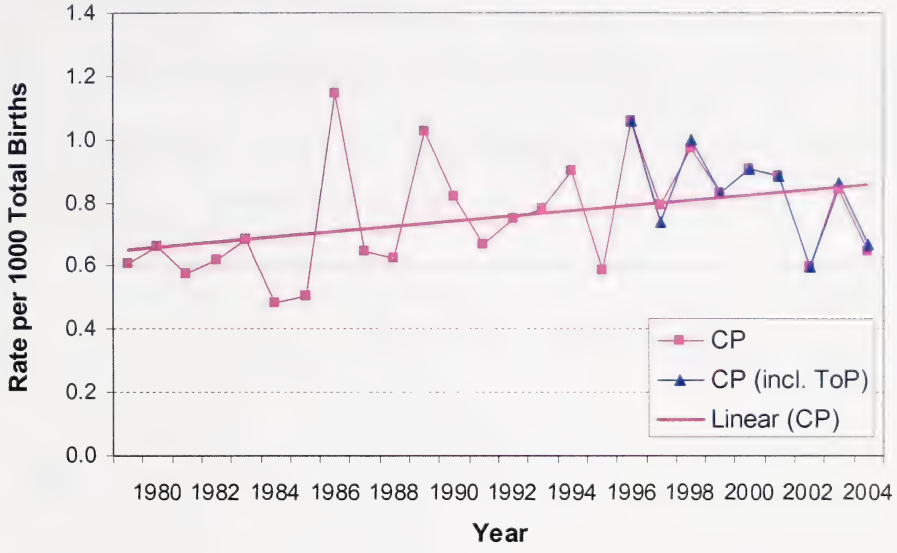


Figure 4.1.5 Cleft Palate with and without ToPs/Foetal Losses, 1980-2005



4.1.3 Abdominal Wall Defects

Abdominal wall defects include mainly gastroschisis and omphalocele. Gastroschisis rates are increasing, a phenomenon that is noted world wide particularly in younger mothers, whereas omphalocele rates have remained stable over the years (**Figure 4.1.6-4.1.8**). Here too we participated in an international study of gastroschisis and associated defects, again co-ordinated by the ICBDSR (see Brit Med J; 332: 423-424. 2006).

Figure 4.1.6 Abdominal Wall Defects (Live births and Stillbirths), 1980-2005

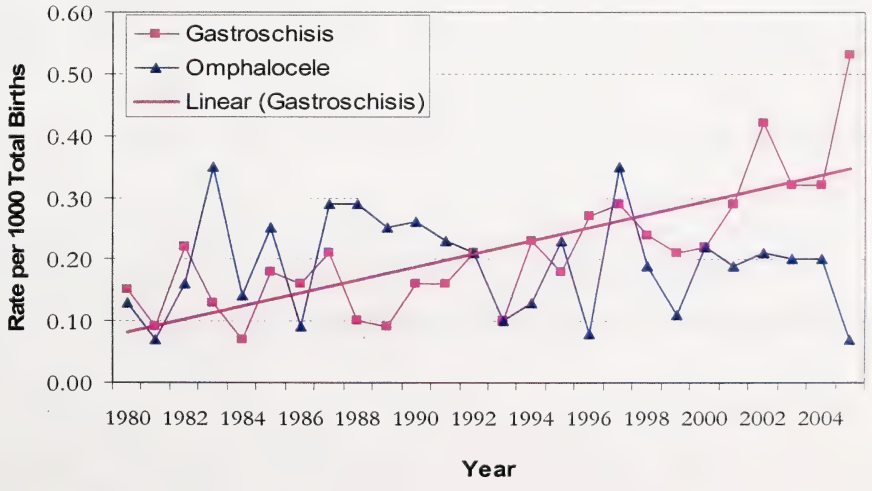


Figure 4.1.7 Gastroschisis with and without ToPs/Foetal Losses, 1980-2005

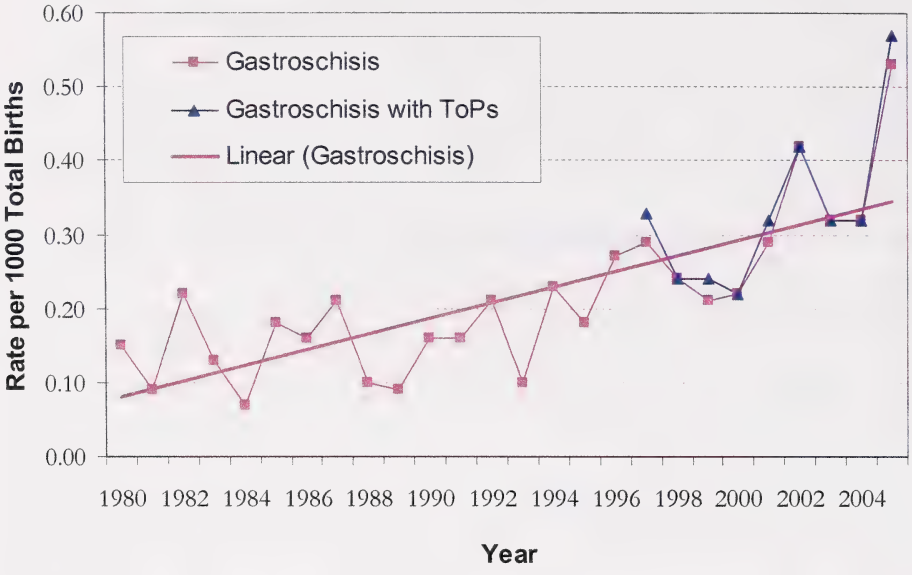
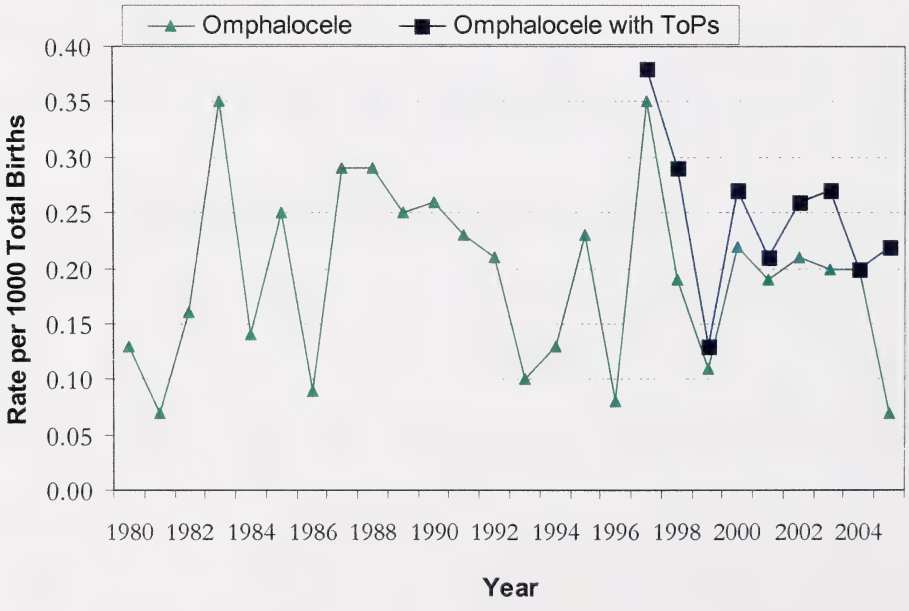


Figure 4.1.8 Omphalocele with and without ToPs/Foetal Losses, 1980-2005



4.1.4 Chromosome Anomalies

From 1980-2005 there were 2285 chromosomal anomalies reported to ACASS. Of these, 1681 or 74% were either Trisomy 13 (Patau Syndrome), Trisomy 18 (Edwards Syndrome) or Trisomy 21 (Down Syndrome). Down Syndrome was by far the most commonly ascertained chromosome anomaly - 77% of the above mentioned group of trisomies and 56% of the total number of chromosome anomalies reported. Sex chromosome anomalies accounted for approximately 8% of the total.

As previously reported, Down Syndrome rates are increasing (**Figure 4.1.9**). In most instances, Down Syndrome is associated with increasing maternal age which is a well reported phenomenon.

Figure 4.1.9 Chromosome Anomalies: Trisomy 13, Trisomy 18, Trisomy 21, 1980-2005

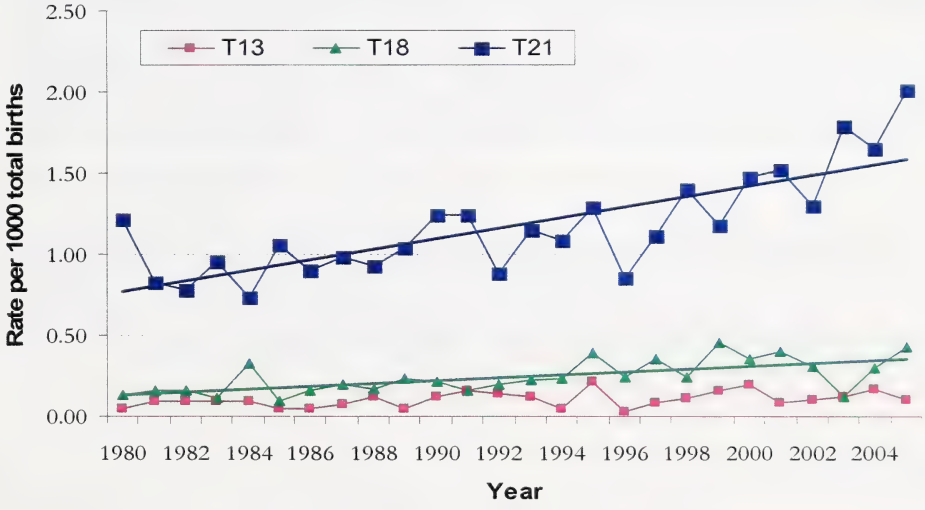
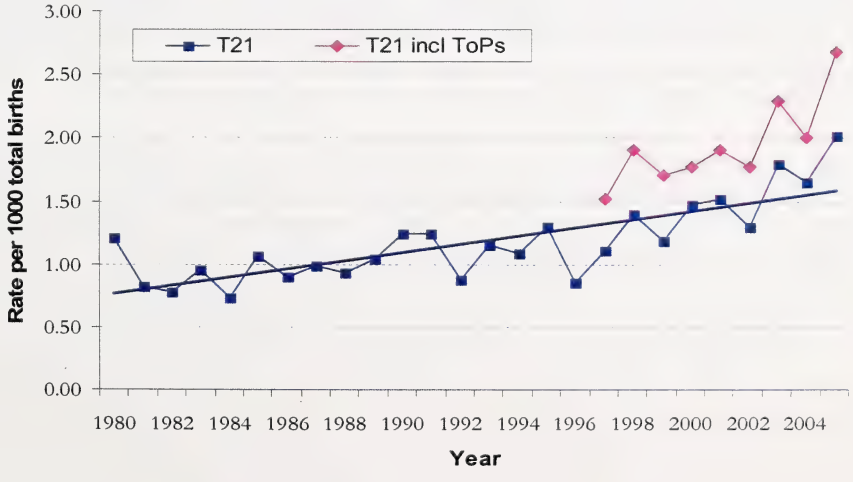


Figure 4.1.10 Down Syndrome with and without ToPs/Foetal Losses, 1980-2005



It is interesting to note that maternal age at delivery is increasing overall in Alberta, in particular the percentage of babies born to women over 35 years of age (**Figure 4.2.1,** page 20).

Terminations of pregnancy (ToPs) do not affect rates of Down Syndrome greatly until we examine births to women over the age of 30 (**Figure 4.1.10, Table 4.2**). As mentioned earlier in the report, ACASS has collected data on ToPs since 1997. Table 4.2 illustrates the rates of Down Syndrome by maternal age for the years 2000 to 2004. The rates including ToPs are in brackets.

Table 4.2 Down Syndrome Rates per 1000 total births by Maternal Age, 2000-2004

Maternal Age	Year				
	2000	2001	2002	2003	2004
<20	0.41 (0.41)	0 (0.43)	0.45 (0.89)	1.86 (1.86)	0.47 (0.47)
20-24	0.53 (0.53)	0.26 (0.40)	0.26 (0.26)	0.87 (0.87)	0.88 (0.88)
25-29	1.32 (1.32)	1.39 (1.39)	0.67 (0.92)	0.97 (0.97)	0.71 (0.78)
30-34	1.78 (1.98)	1.52 (1.71)	1.37 (1.37)	1.30 (1.74)	1.19 (1.53)
35-39	2.37 (3.45)	3.01 (4.29)	4.20 (5.88)	4.42 (5.63)	5.81 (6.81)
40-44	6.46 (11.63)	11.35 (16.39)	4.94 (12.35)	11.33 (20.60)	7.29 (11.46)
≥ 45	0 (0)	0 (0)	0 (0)	22.73 (22.73)	0 (0)

Infants with Down Syndrome often have associated anomalies. ACASS does not code minor anomalies associated with Down Syndrome such as single palmar crease, upslanting palpebral fissures, and increased space between the great and second toes. However, most other malformations, if mentioned on the ascertainment documents, are entered routinely into the database. Anomalies such as patent ductus arteriosus, undescended testes and lung hypoplasia are not coded if the infant was born prematurely or weighed less than 2500 grams. This is consistent with our general coding policies.

4.1.5 Limb Reductions

Limb reduction defects are increasing and will be the focus of more in-depth scrutiny in the future (**Figure 4.1.11**). There are a number of possible explanations aside from a true increase in the rates. Due to computer limitations in the years 1980 – 1996 inclusive we were able to code only 6 anomalies per case. Since 1997, we have been able to code an unlimited number of defects per case. The cases will be reviewed to try to determine whether the upward trend is due to improved ascertainment, different coding procedures or whether in fact the rate has truly increased.

ACASS is also participating in an international study, co-ordinated through the ICBDSR, on the epidemiology of very rare defects among which are some of the more uncommon limb reduction defects such as true phocomelia (absence of all limb bones proximal to the hand or foot, which attaches directly to the trunk) and amelia (complete absence of one or more limbs).

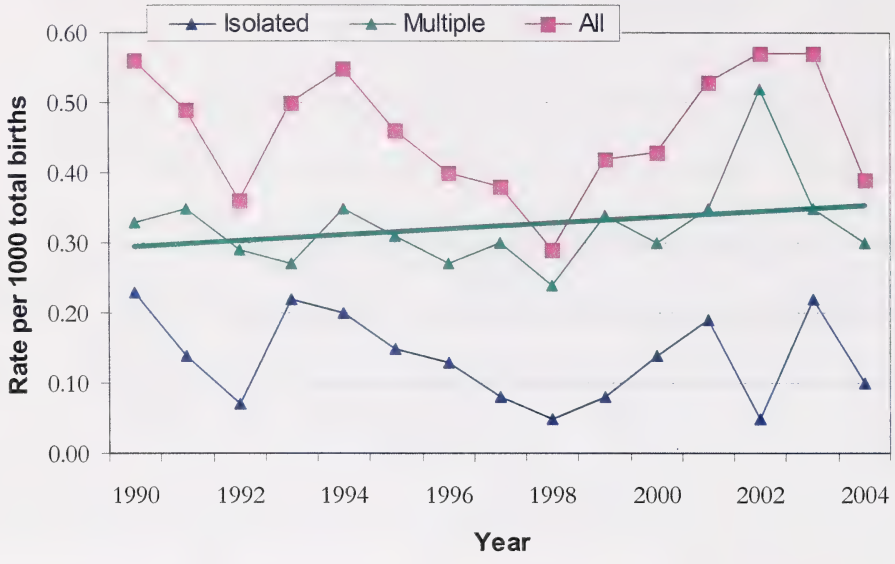
Figure 4.1.11. Limb Reduction Defects with and without ToPs/Foetal Losses, 1980-2005

4.1.6 Anorectal atresia/stenosis

Although anorectal anomalies plus large intestine anomalies appear to be increasing (they are lumped together in the basic ICD code), when one examines the anorectal portion of the category alone, the rates have been stable over the past 15 years (1990-2004). Initially the rates appeared to increase in 1998 in both Alberta and in the Canadian National data which prompted a detailed survey of the Alberta data. A manuscript has been accepted for publication by the Journal of Pediatric Surgery (in press) with the results of the survey. While rates do fluctuate between 3 and 5 per 1000 total births, there is no overall trend in the study period for all cases (isolated and multiple combined) and isolated cases but perhaps a slight increasing trend for multiples although not statistically significant (**Figure 4.1.12**). The results do indicate however that a substantial number of cases belong in the multiple congenital anomaly category VATER/VACTERL (**Figure 4.1.12**).

Our prevalence rates are very comparable to two other large population studies, one from British Columbia and the other from the EUROCAT registries, with frequencies in the 1/2200 – 1/2500 range.

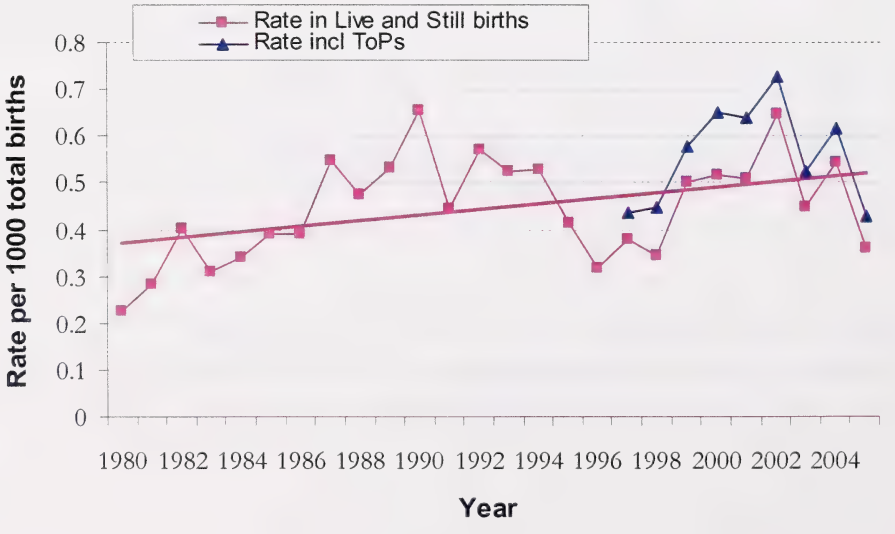
Figure 4.1.12 Anorectal Atresia/Stenosis, Isolated and Multiple Anomalies, 1990-2004



4.1.7 Renal Agenesis/Hypoplasia

The rates of renal agenesis appear to be increasing (**Figure 4.1.13**). This is likely due to improved diagnostic capabilities hence better ascertainment.

Figure 4.1.13 Renal Agenesis/Hypoplasia, 1980-2005



4.1.8 Summary

ACASS reviews anomalies that have been entered into the database on a regular basis. As is evident from the review of anorectal atresia and neural tube defects, a detailed study of some individual items aids in the assessment and maintenance of the data quality. With intensive review, some cases might be re-assigned, re-coded or discarded altogether from the database. This continuing review might explain some discrepancies in the data from earlier reports.

4.2 Birth Prevalence by Maternal Age

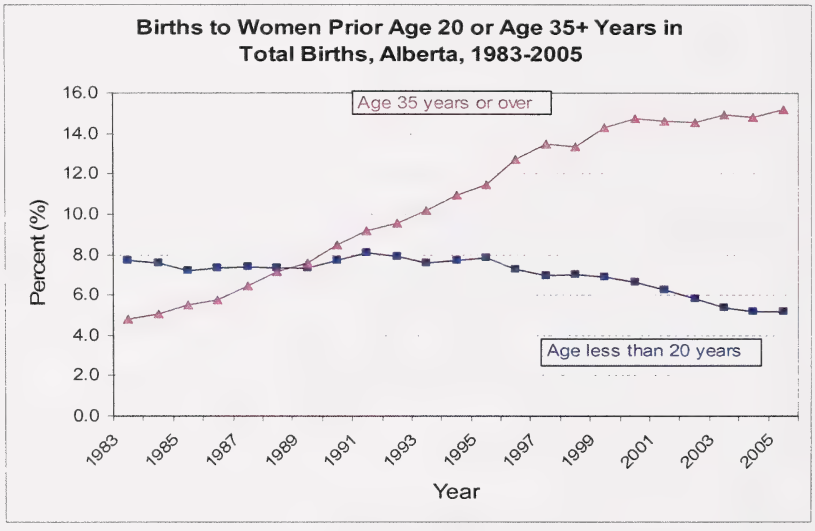
4.2.1 Background

Maternal age is a major risk indicator for congenital anomalies. Women giving birth over the age of 34 or under age 20 have an increased risk for certain congenital anomalies (ACASS report, 1999; Baird et al., 1991; Reefhuis et al., 2004; Bianca et al., 2005). Maternal age is also an indicator for prenatal screening for Down syndrome (Serra-Prat, et al., 1998; Saltvedt et al., 2005).

Women giving birth over the age of 34 have an increased risk for Down syndrome, other chromosomal anomalies, musculoskeletal anomalies, heart defects, and genital organ anomalies (Baird et al., 1991; Reefhuis et al., 2004; Bianca et al., 2005). Teenage pregnancies are associated with an increased risk for some anomalies of the nervous, circulatory, and musculoskeletal systems (Reefhuis et al., 2004; Vieira et al., 2005; Cleary-Goldman et al., 2005).

4.2.2 Time Trends (see Fig. 4.2.1)

Fig.4.2.1

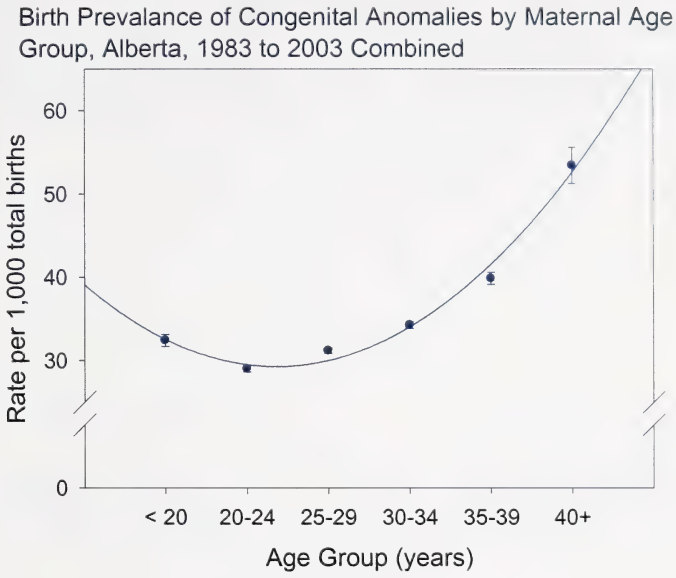


The proportion of births to women 35 years of age or over has increased in Alberta, from 4.8% in 1983 to 15.2% in 2005 **(Figure 4.2.1)**. In contrast, the proportion of births to women under 20 years of age has decreased, from 7.7% in 1983 to 5.2% in 2005.

4.2.3 Age Effects (see Table 4.2.1)

Overall, the birth prevalence rate for all congenital anomalies follows a "J" shape pattern in Alberta (Figure 4.2.2). Women giving birth between the ages of 20 and 24 years have the lowest congenital anomaly rate (29.0 per 1,000 total births). Compared to this group, women in the other age groups show an increased congenital anomaly rate (Fig. 4.2.2).

Fig.4.2.2



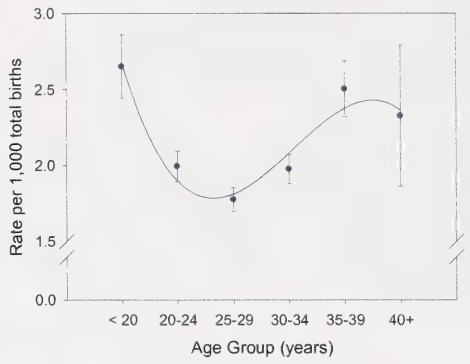
Looking at specific anomaly groups, women giving birth at age 35 years or over have an increased birth prevalence rate for the following groups (**Fig.4.2.3.1, Fig. 4.2.3.2)**: Down syndrome, ventricular septal defects, nervous system defects, cleft palate, renal dysgenesis or agenesis, congenital hip dislocation, and abdominal wall defects (excluding gastroschisis). Other chromosomal anomalies, other circulatory defects, urinary obstructive defects, hypospadias and epispadias also have an increased birth prevalence in women 35 years or over (data not shown).

Women giving birth before age 20 have an increased birth prevalence rate for nervous system defects, ventricular septal defects, cleft lip +/- cleft palate, and gastroschisis (**Fig. 4.2.3.1, 4.2.3.2**).

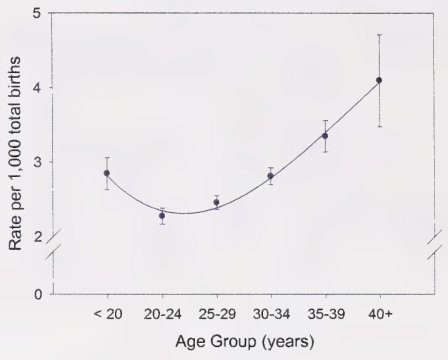
Detailed data on birth prevalence of selected congenital anomalies are presented in **Table 4.2.1** (page 25).

Fig. 4.2.3.1 Birth Prevalence of Selected Congenital Anomalies by Maternal Age Group Alberta, 1983-2003 Combined

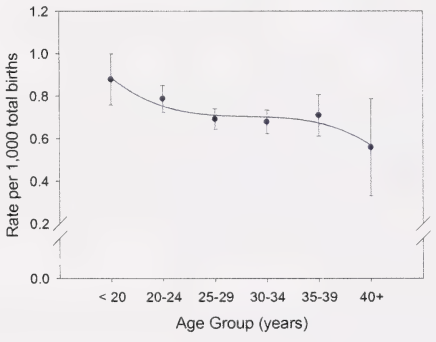
Nervous system anomalies
ICD-9=740-742, ICD-10=Q00-Q07



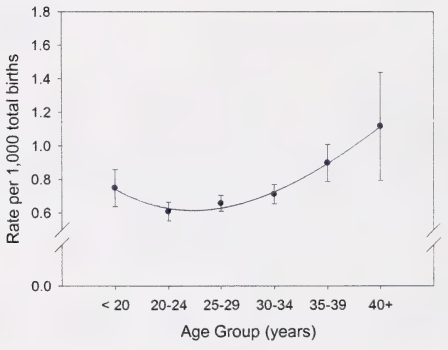
Ventricular septal defects
ICD-9=745.4, ICD-10=Q21.0,Q21.8



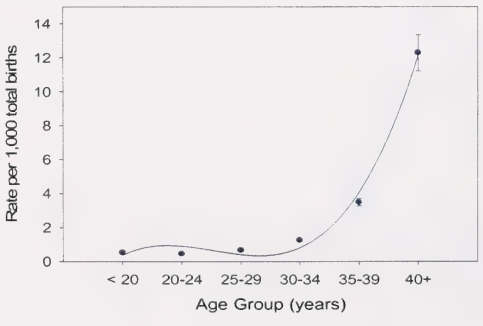
Neural Tube Defects (NTDs)
ICD-9=740-741,720.0, ICD-10=Q00,Q01,Q05



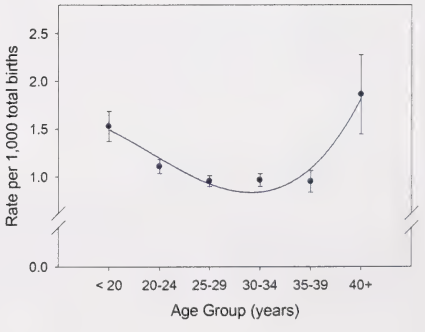
Cleft palate
ICD-9=749.0, ICD-10=Q35



Down syndrome
ICD-9=758.0, ICD-10=Q90



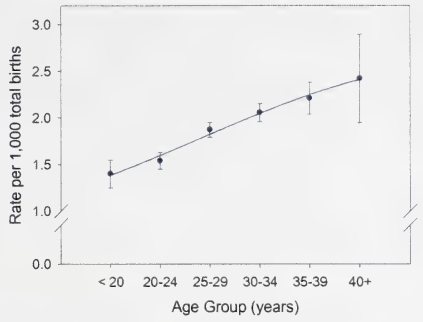
Cleft lip +/- cleft palate
ICD-9=749.1-749.2, ICD-10=Q36-Q37



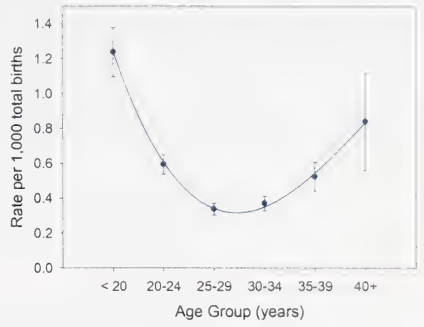
Source: 1. Vital Statistics, Birth File, Department of Government Services, March 2006 release.
2. Alberta Congenital Anomalies Surveillance System (ACASS), July 2006 release.

Fig. 4.2.3.2 Birth Prevalence of Selected Congenital Anomalies by Maternal Age Group, Alberta, 1983-2003 Combined

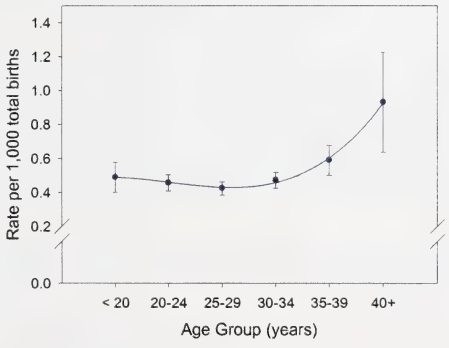
Musculoskeletal - hip dislocation
ICD-9=754.3, ICD-10=Q65



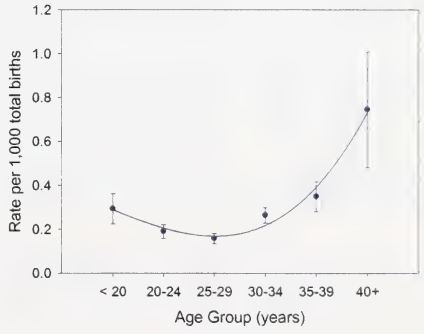
Abdominal wall defects
ICD-9=756.7, ICD-10=Q79.2-Q79.5



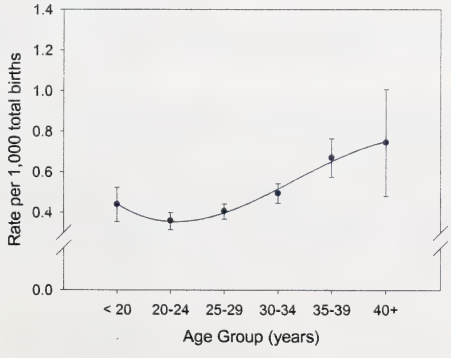
Cystic kidney disease
ICD-9=753.1, ICD-10=Q61



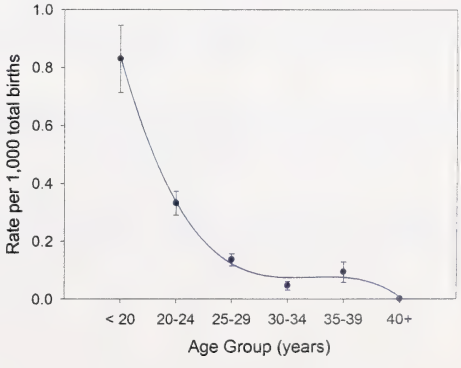
Omphalocele
ICD-9=756.70, ICD-10=Q79.2



Renal dysgenesis or agenesis
ICD-9=753.0, ICD-10=Q60



Gastroschisis
ICD-9=756.71, ICD-10=Q79.3



Source: 1. Vital Statistics, Birth File, Department of Government Services, March 2006 release.
2. Alberta Congenital Anomalies Surveillance System (ACASS), July 2006 release.

4.2.4 Limitations and Methodology Notes

A case refers to a baby with a given congenital anomaly identified by the 1st birthday.

Cases include congenital anomalies in live births, stillbirths and terminated pregnancies.

One baby may have more than one anomaly, and thus may be counted in more than one category.

Some specific groups of congenital anomalies were combined to increase the reliability of the birth prevalence rate, such as nervous system anomalies, hypospadias and epispadias.

A small number of cases in 2005 may be missed from reporting at the time of release of the ACASS data in July 2006.

A small number of cases did not include maternal age at delivery, especially cases prior to 1986.

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Bianca S, Ingegnosi C, Ettore G. Maternal age and hypospadias. Acta Obstet Gynecol Scand 2005; 84(4):410.

Table 4.2.1 Number and Birth Prevalence of Selected Congenital Anomalies by Maternal Age, Alberta, 1983 to 2003 Combined

Selected anomalies	Maternal Age at Delivery (year)						All Ages
	< 20	20-24	25-29	30-34	35-39	40+	
1. Total congenital anomalies							
Number of cases	1,992	5,669	9,150	7,419	2,976	574	31,992
Rate (per 1,000 total births)	32.4	29.0	31.2	34.2	39.8	53.4	37.5
Standard Error (SE)	0.71	0.38	0.32	0.39	0.72	2.17	0.21
2. Neural tube defects (NTDs)							
Number of cases	54	154	203	147	53	6	729
Rate (per 1,000 total births)	0.9	0.8	0.7	0.7	0.7	0.6	0.9
Standard Error (SE)	0.12	0.06	0.05	0.06	0.10	0.23	0.03
3. Ventricular septal defects (VSDs)							
Number of cases	175	445	721	609	250	44	2,507
Rate (per 1,000 total births)	2.8	2.3	2.5	2.8	3.3	4.1	2.9
Standard Error (SE)	0.21	0.11	0.09	0.11	0.21	0.62	0.06
4. Cleft lip +/- cleft palate							
Number of cases	94	217	280	209	71	20	1,008
Rate (per 1,000 total births)	1.5	1.1	1.0	1.0	1.0	1.9	1.2
Standard Error (SE)	0.16	0.08	0.06	0.07	0.11	0.42	0.04
5. Cleft palate							
Number of cases	46	119	193	154	67	12	659
Rate (per 1,000 total births)	0.7	0.6	0.7	0.7	0.9	1.1	0.8
Standard Error (SE)	0.11	0.06	0.05	0.06	0.11	0.32	0.03
6. Down syndrome							
Number of cases	33	91	199	274	261	132	1,088
Rate (per 1,000 total births)	0.5	0.5	0.7	1.3	3.5	12.3	1.3
Standard Error (SE)	0.09	0.05	0.05	0.08	0.22	1.06	0.04
7. Cystic kidney disease							
Number of cases	30	89	124	102	44	10	430
Rate (per 1,000 total births)	0.5	0.5	0.4	0.5	0.6	0.9	0.5
Standard Error (SE)	0.09	0.05	0.04	0.05	0.09	0.29	0.02
8. Renal dysgenesis or agenesis							
Number of cases	27	70	119	107	50	8	425
Rate (per 1,000 total births)	0.4	0.4	0.4	0.5	0.7	0.7	0.5
Standard Error (SE)	0.08	0.04	0.04	0.05	0.09	0.26	0.02
9. Congenital hip dislocation							
Number of cases	86	301	549	445	165	26	1,871
Rate (per 1,000 total births)	1.4	1.5	1.9	2.1	2.2	2.4	2.2
Standard Error (SE)	0.15	0.09	0.08	0.10	0.17	0.47	0.05
10. Gastroschisis							
Number of cases	51	65	40	10	7	0	173
Rate (per 1,000 total births)	0.8	0.3	0.1	0.0	0.1	0.0	0.2
Standard Error (SE)	0.12	0.04	0.02	0.01	0.04	0.00	0.02
11. Omphalocele							
Number of cases	18	37	46	57	26	8	193
Rate (per 1,000 total births)	0.3	0.2	0.2	0.3	0.3	0.7	0.2
Standard Error (SE)	0.07	0.03	0.02	0.03	0.07	0.26	0.02
12. Abdominal wall defects							
Number of cases	76	116	99	80	39	9	424
Rate (per 1,000 total births)	1.2	0.6	0.3	0.4	0.5	0.8	0.5
Standard Error (SE)	0.14	0.06	0.03	0.04	0.08	0.28	0.02

Source: 1. Vital Statistics, Birth File, Department of Government Services, March 2006 release.
2. Alberta Congenital Anomalies Surveillance System (ACASS), July 2006 release.

Notes: Data include Alberta residents only.
Totals for Alberta include data of unknown maternal age.
Data may differ from previously published data due to differences in definitions and dates of data release.

4.3 Birth Prevalence by Birth Weight

4.3.1 Background

Low birth weight refers to a birth weight less than 2500 grams. Extremely low birth weight (ELBW) refers to birth weight less than 1,000 grams, and very low birth weight (VLBW) to less than 1500 grams.

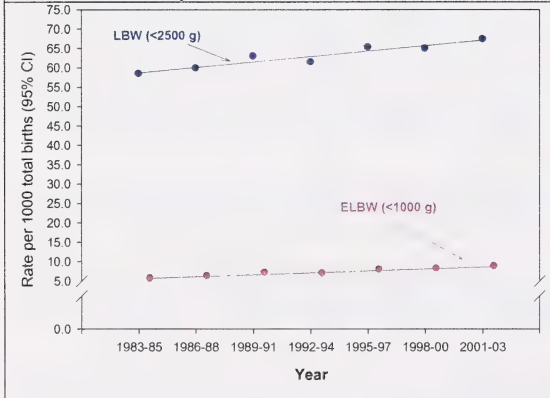
Low birth weight is correlated with fetal, neonatal and long term complications, including physical, cognitive, and behavioral impairments (Anderson et al., 2003; Jarvis et al., 2003), as well as fetal and infant mortality (Chen et al., 1998). The risk is particularly high when the birth weight is less than 1000 grams.

Congenital anomalies are closely related to low birth weight. About 19% of infants with congenital anomalies in Alberta were low birth weight. In births to mothers over 34 years of the age, this proportion was higher (24%). The birth prevalence of congenital anomalies may vary by birth weight.

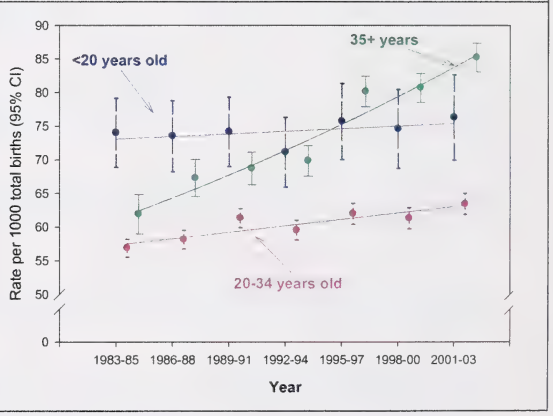
4.3.2 Time Trends of Low Birth Weight (see Fig. 4.3.1)

Fig. 4.3.1

Low Birth Weight and Extreme Low Birth Weight Rate by 3-Year Period, 1983-2003



Low Birth Weight Rate by Maternal Age and 3-Year Period, 1983-2003

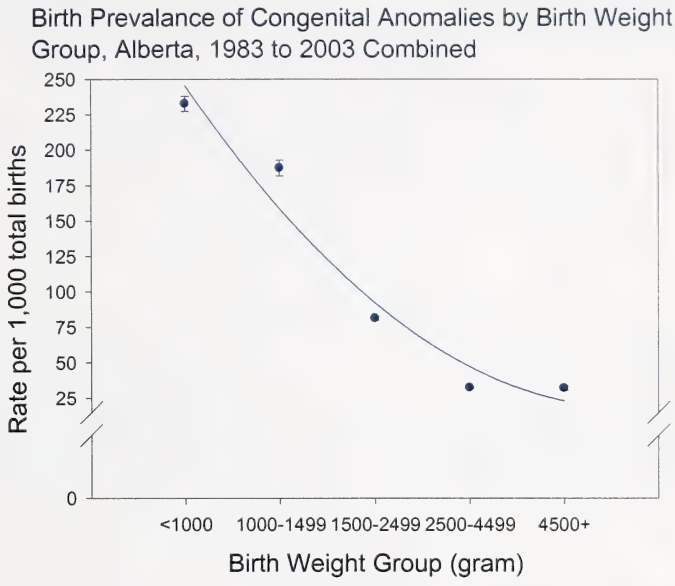


The rate of low birth weight babies has increased in Alberta, from 58.5 per 1000 total births in 1983-1985 to 67.4 in 2001-2003. The extreme low birth weight (ELBW) rate also increased from 5.7 per 1000 total births in 1983-1985 to 8.7 in 2001-2003 (**Figure 4.3.1**). The greatest increase occurred among births to mothers over 34 years of age (**Figure 4.3.1**).

In 2001-2003, there were 7,826 low birth weight babies born in Alberta. Of these, 1,028 (13.1%) were extremely low birth weight (ELBW), and 1,757 (22.4%) were very low birth weight (VLBW). About 19% of the low birth weight babies were born to mothers over 34 years of age, 13% had congenital anomalies, and 7% were stillbirths.

4.3.3 Birth Prevalence by Birth Weight Group (see Table 4.3.1, 4.3.2)

Fig. 4.3.2



Overall, the birth prevalence of all congenital anomalies was higher for low birth weight babies in Alberta in 1983-2003 combined, ranging from 232.7 per 1000 total births for ELBW babies, to 187.3 for 1000-1499 gram babies, and to 81.2 for 1500-2499 gram babies. The rate was 32.4 per 1000 total births for babies with birth weight between 2500 and 4499 grams (**Fig. 4.3.2**).

Anomalies increasing with decreasing birth weight: In general, most diagnostic groups of congenital anomalies showed increasing birth prevalence with decreasing birth weight in Alberta from 1983 to 2003 (**Fig.4.3.3.1, Fig.4.3.3.2**).

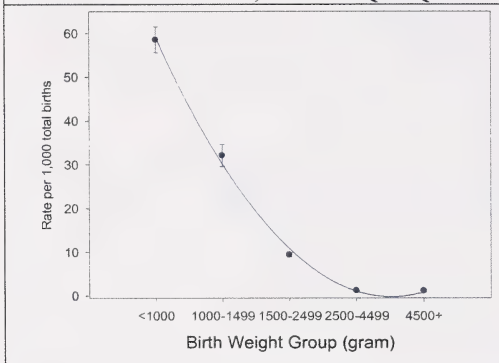
Anomalies with lower birth prevalence in ELBW group: The birth prevalence rate was NOT higher in ELBW (<1000 g) babies than 1000-1499 gram babies for the following groups of congenital anomalies: ventricular septal defects (**Fig.4.3.3.1**), genital organ anomalies (i.e., hypospadias and epispadias), diaphragmatic hernia, gastroschisis (**Fig. 4.3.3.3**), and eye anomalies (data not shown).

Anomalies decreasing with decreasing birth weight: The birth prevalence rate decreased with decreasing birth weight for congenital hip dislocation in Alberta from 1983 to 2003 (**Figs. 4.3.3.3**).

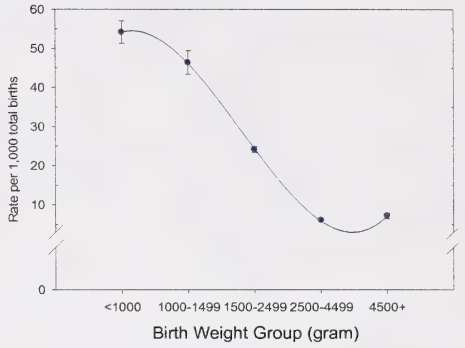
Detailed data on birth prevalence of selected congenital anomalies are presented in **Table 4.3.1 (page 32).**

Fig. 4.3.3.1 Birth Prevalence of Selected Congenital Anomalies by Birth Weight Group Alberta, 1983-2003 Combined

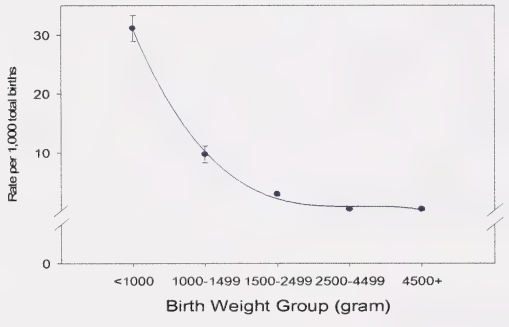
Nervous system anomalies
ICD-9=740-742, ICD-10=Q00-Q07



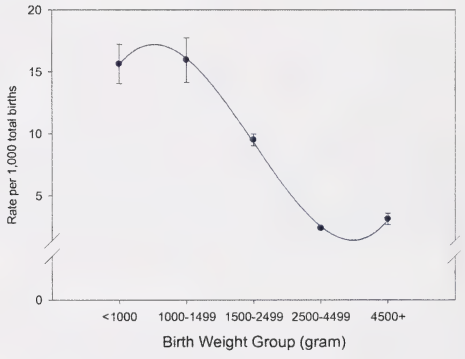
Circulatory system anomalies
ICD-9=745-747, ICD-10=Q20-Q28



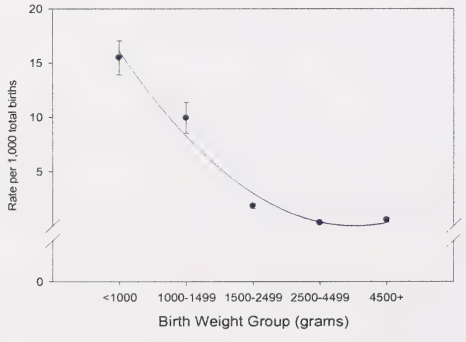
Neural Tube Defects (NTDs)
ICD-9=740-741,720.0, ICD-10=Q00,Q01,Q05



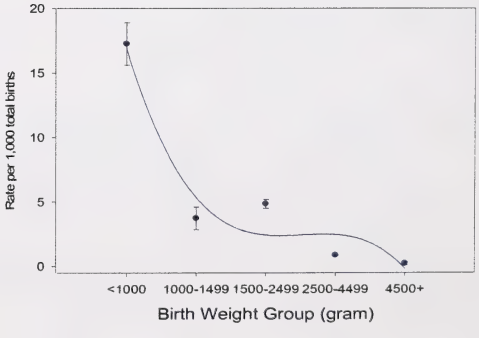
Ventricular septal defect
ICD-9=745.4, ICD-10=Q21.0, Q21.8



Hydrocephalus
ICD-9=742.3, ICD-10=Q03



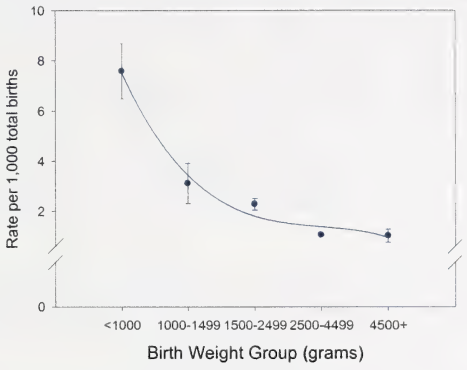
Down syndrome
ICD-9=758.0, ICD-10=Q90



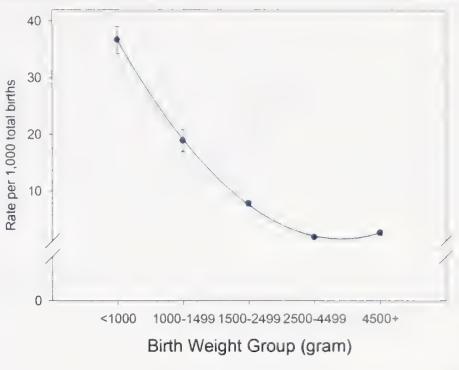
Source: 1. Vital Statistics, Birth File, Department of Government Services, March 2006 release.
2. Alberta Congenital Anomalies Surveillance System (ACASS), July 2006 release.

Fig. 4.3.3.2 Birth Prevalence of Selected Congenital Anomalies by Birth Weight Group Alberta, 1983-2003 Combined

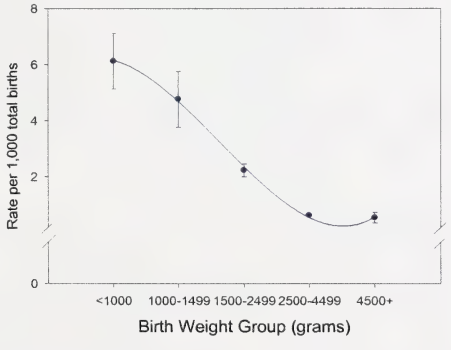
Digestive - Cleft lip +/- cleft palate
ICD-9=749.1-749.2, ICD-10=Q36-Q37



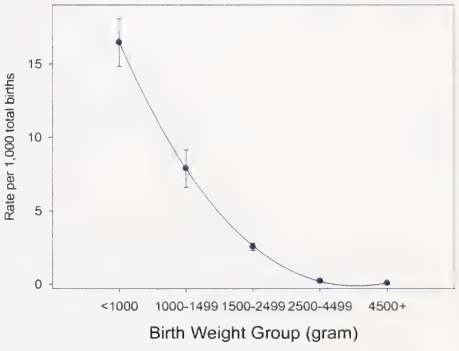
Urinary system anomalies
ICD-9=753, ICD-10=Q60-Q64



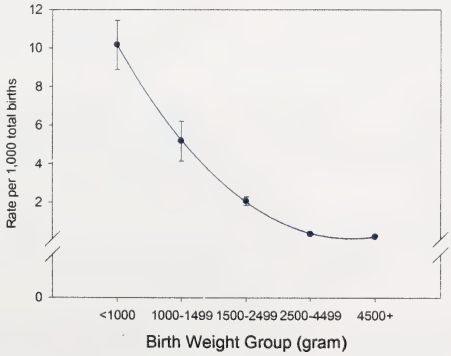
Digestive - Cleft palate
ICD-9=749.0, ICD-10=Q35



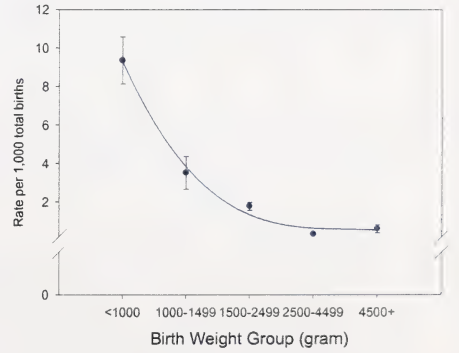
Renal dysgenesis or agenesis
ICD-9=753.0, ICD-10=Q60



Rectal/large intestinal atresia/stenosis
ICD-9=751.2, ICD-10=Q42



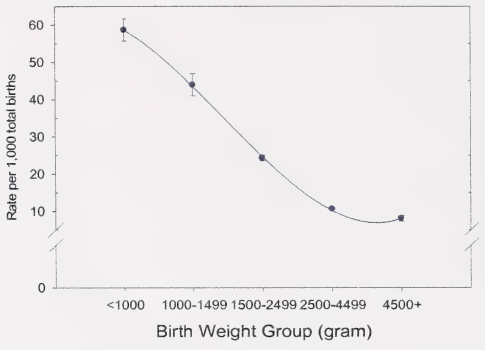
Cystic kidney disease
ICD-9=753.1, ICD-10=Q61



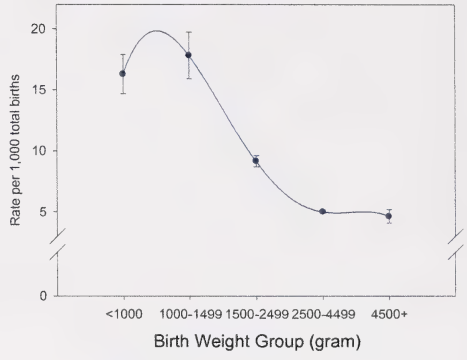
Source: 1. Vital Statistics, Birth File, Department of Government Services, March 2006 release.
2. Alberta Congenital Anomalies Surveillance System (ACASS), July 2006 release.

Fig. 4.3.3.3 Birth Prevalence of Selected Congenital Anomalies by Birth Weight Group Alberta, 1983-2003 Combined

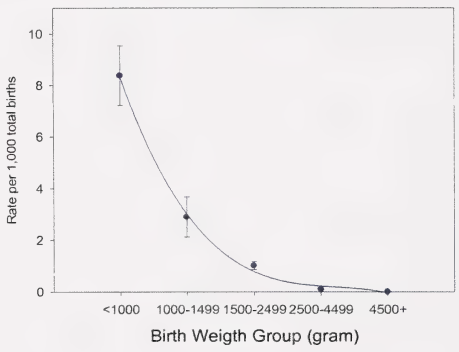
Musculoskeletal anomalies
ICD-9=754-756, ICD-10=Q65-Q79



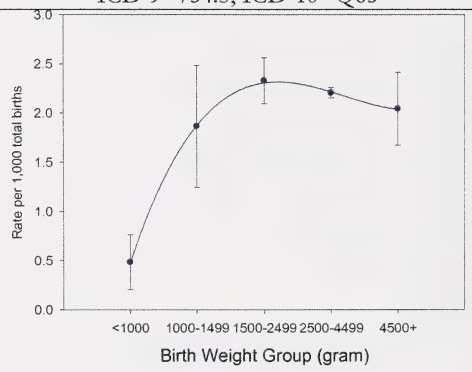
Genital organ anomalies
ICD-9=752, ICD-10=Q50-Q56



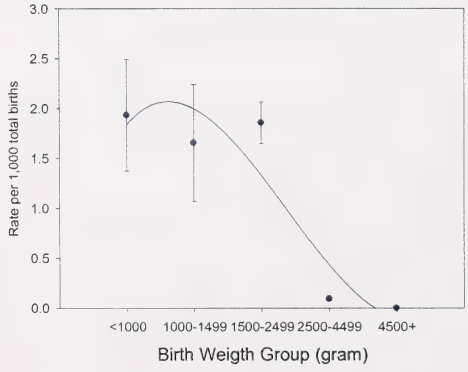
Omphalocele
ICD-9=756.70, ICD-10=Q79.2



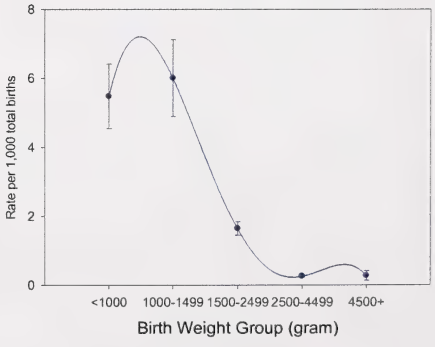
Musculoskeletal - hip dislocation
ICD-9=754.3, ICD-10=Q65



Gastroschisis
ICD-9=756.71, ICD-10=Q79.3



Musculoskeletal - diaphragmatic hernia
ICD-9=756.6, ICD-10=Q79.0-Q79.1



Source: 1. Vital Statistics, Birth File, Department of Government Services, March 2006 release.
2. Alberta Congenital Anomalies Surveillance System (ACASS), July 2006 release.

4.3.4 Limitations and Methodology Notes

The low birth weight rate is defined as the number of low birth weight births per 1,000 total births (live births plus stillbirths).

Cases included congenital anomalies in live births, stillbirths and terminated pregnancies.

Low birth weight cases may be preterm (born at < 37 weeks of gestation). The proportion of preterm cases in 2001-2003 was 98.9%, 98.5%, 71.2%, and 6.8% for the <1000, 1000-1499, 1500-2499, and 2500-4499 gram birth weight groups, respectively.

The birth weight-specific birth prevalence rate of congenital anomalies is expressed as the number of anomalies per 1,000 total births (live births plus stillbirths) in each birth weight group (<1000, 1000-1499, 1500-2499, 2500-4499, 4500+ grams).

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Table 4.3.1 Number and Birth Prevalence of Selected Congenital Anomalies by Birth Weight Group, Alberta, 1983 to 2003 Combined

Selected anomalies	Birth Weight Group (gram)					
	<1000	1000-1499	1500-2499	2500-4499	4500+	All Groups
1 Total congenital anomalies						
Number of cases	1,443	904	3,452	25,390	469	31,992
Rate (per 1,000 total births)	232.7	187.3	81.2	32.4	31.9	37.5
Standard Error (SE)	5.37	5.62	1.32	0.20	1.45	0.21
2 Neural tube defects (NTDs)						
Number of cases	193	47	126	322	6	729
Rate (per 1,000 total births)	31.1	9.7	3.0	0.4	0.4	0.9
Standard Error (SE)	2.20	1.41	0.26	0.02	0.17	0.03
3 Congenital hydrocephalus						
Number of cases	96	48	78	223	8	456
Rate (per 1,000 total births)	15.5	9.9	1.8	0.3	0.5	0.5
Standard Error (SE)	1.57	1.43	0.21	0.02	0.19	0.03
4. Ventricular septal defects (VSDs)						
Number of cases	97	77	404	1,872	46	2,507
Rate (per 1,000 total births)	15.6	16.0	9.5	2.4	3.1	2.9
Standard Error (SE)	1.58	1.80	0.47	0.06	0.46	0.06
5. Cleft lip +/- cleft palate						
Number of cases	47	15	97	825	15	1,008
Rate (per 1,000 total births)	7.6	3.1	2.3	1.1	1.0	1.2
Standard Error (SE)	1.10	0.80	0.23	0.04	0.26	0.04
6. Cleft Palate						
Number of cases	38	23	95	493	8	659
Rate (per 1,000 total births)	6.1	4.8	2.2	0.6	0.5	0.8
Standard Error (SE)	0.99	0.99	0.23	0.03	0.19	0.03
7. Down syndrome						
Number of cases	107	18	206	668	3	1,088
Rate (per 1,000 total births)	17.3	3.7	4.8	0.9	0.2	1.3
Standard Error (SE)	1.65	0.88	0.34	0.03	0.12	0.04
8. Cystic kidney disease						
Number of cases	58	17	76	265	9	430
Rate (per 1,000 total births)	9.4	3.5	1.8	0.3	0.6	0.5
Standard Error (SE)	1.22	0.85	0.20	0.02	0.20	0.02
9. Renal dysgenesis or agenesis						
Number of cases	102	38	108	163	1	425
Rate (per 1,000 total births)	16.4	7.9	2.5	0.2	0.1	0.5
Standard Error (SE)	1.61	1.27	0.24	0.02	0.07	0.02
10 Rectal/large intestinal atresia/stenosis						
Number of cases	63	25	88	270	3	457
Rate (per 1,000 total births)	10.2	5.2	2.1	0.3	0.2	0.5
Standard Error (SE)	1.27	1.03	0.22	0.02	0.12	0.03
11 Omphalocele						
Number of cases	52	14	43	73	0	193
Rate (per 1,000 total births)	8.4	2.9	1.0	0.1	0.0	0.2
Standard Error (SE)	1.16	0.77	0.15	0.01	0.00	0.02
12. Gastroschisis						
Number of cases	12	8	79	72	0	173
Rate (per 1,000 total births)	1.9	1.7	1.9	0.1	0.0	0.2
Standard Error (SE)	0.56	0.59	0.21	0.01	0.00	0.02

Source: 1. Vital Statistics, Birth File, Department of Government Services, March 2006 release.
2. Alberta Congenital Anomalies Surveillance System (ACASS), July 2006 release.

Notes: Data include Alberta residents only.
Totals for Alberta include data of unknown birth weight.
Data may differ from previously published data due to differences in definitions and dates of data release.

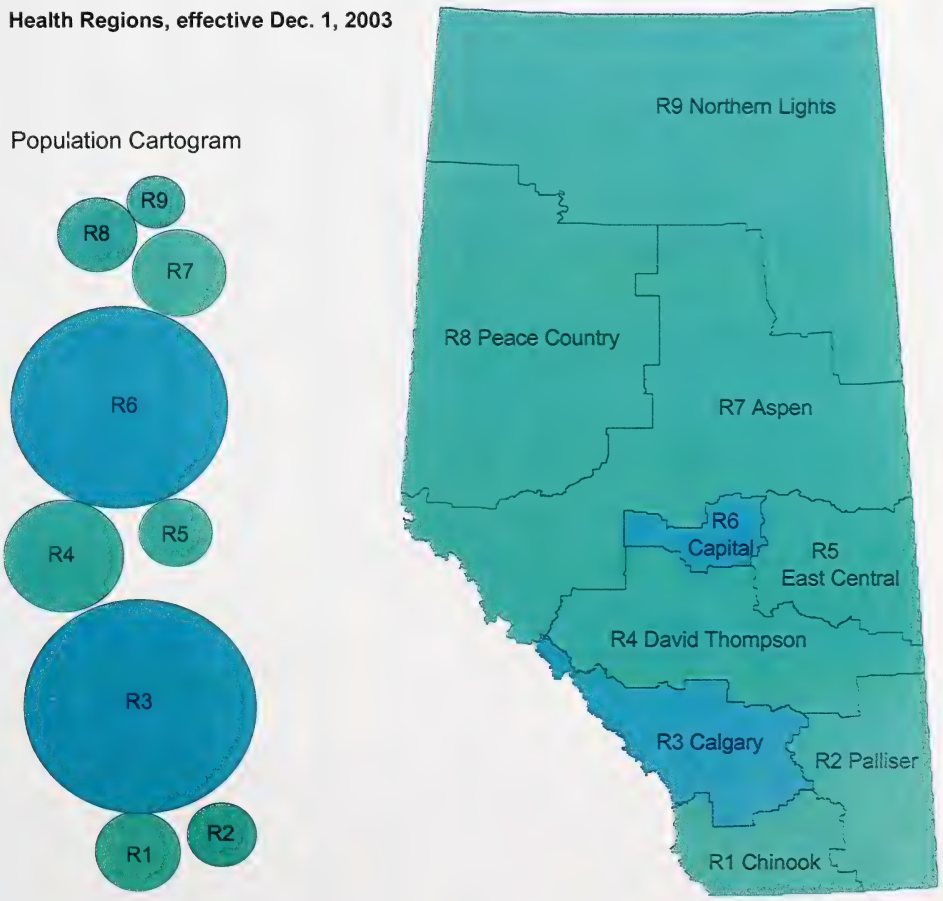
4.4 Birth Prevalence – Regional Data

4.4.1 Background

The birth prevalence of congenital anomalies by health region has been reported in earlier surveillance reports (Child Health Surveillance Report, 2006; Health Trends, 2001, 2006) and this section is an update of the regional congenital anomaly data in Alberta for 2003-2005 combined.

The regional variation in congenital anomalies may in part be related to regional variations in case ascertainment and reporting and differences in demographics, risk factors (i.e., maternal, behavioural), access to prenatal care and maternal conditions across regions. The Calgary Health Region had a higher rate of low birth weight, SGA, preterm births and a higher proportion of births to women over 34 years of age. Regional Health Authorities (RHA) 2, 7, 8, and 9 had higher rates of maternal obesity, smoking and alcohol consumption during pregnancy (Reproductive Health Report, 2004, 2006).

The Map of Alberta with Regional Health Authorities and Population Size



4.4.2 Regional Data (see Table 4.4.1)

Table 4.4.1 presents the birth prevalence of selected congenital anomalies by health region, 2003-2005 combined.

The rate of all congenital anomalies was higher than the provincial average in RHA 3, with a rate of 49.3, or 2,203 cases for 2003 to 2005 combined. This increase may in part be due to better case ascertainment and reporting in the Calgary Health Region.

The rate of Neural Tube Defects (NTDs) was lower than the provincial average in RHA 1 and higher in RHAs 5, 7 and 8. The rate of eye anomalies was slightly higher than the provincial average in RHA 2. For ventricular septal defects, the rate was higher in RHAs 1, 3, and 8. The rate of cleft lip +/- cleft palate was lower in RHAs 1, 5, and 9. The rate of hypospadias and epispadias was higher in RHA 3 and lower in RHA 9. For Down's syndrome, the rate was higher than the provincial average for RHAs 3 and 6 and lower in RHA 2. The number of cases for other groups of congenital anomalies is too small for meaningful comparisons.

4.4.3 Limitations and Methodology Notes

Improved diagnosis, reporting, and surveillance, increasing maternal age, and inclusion of pregnancy terminations for fetal anomalies all likely contributed to the increasing rate of congenital anomalies over time.

Many aspects of reporting of congenital anomalies may not be consistent across regions over time, including reporting of fetal anomalies and diagnosis of minor anomalies. These differences could contribute to variations in rates across regions.

Table 4.4.1 Number and Birth Prevalence of Selected Congenital Anomalies by Residence RHA, Alberta, 2003-2005

Selected anomalies	R1-Chinook	R2 - Palliser	R3 - Calgary	R4 - David Thompson	R5 - East Central	R6 - Capital	R7 - Aspen	R8 - Peace Country	R9 - Northern Lights	Alberta
1. All congenital anomalies										
Number of cases	233	163	2,208	342	84	1,119	214	191	84	4,654
Rate (per 1,000 total births)	37.1	41.7	49.3	30.0	30.1	31.1	29.7	31.3	20.8	38.0
Standard Error (SE)	2.38	3.20	1.02	1.60	3.24	0.92	2.00	2.23	2.24	0.55
2 Neural tube defects (NTDs)										
Number of cases	1	2	27	10	3	30	8	6	3	91
Rate (per 1,000 total births)	0.2	0.5	0.6	0.9	1.1	0.8	1.1	1.0	0.7	0.7
Standard Error (SE)	0.16	0.36	0.12	0.28	0.62	0.15	0.39	0.40	0.43	0.08
3. Eye anomalies										
Number of cases	6	6	39	3	2	13	4	3	4	81
Rate (per 1,000 total births)	1.0	1.5	0.9	0.3	0.7	0.4	0.6	0.5	1.0	0.7
Standard Error (SE)	0.39	0.63	0.14	0.15	0.51	0.10	0.28	0.28	0.49	0.07
4. Ventricular septal defects (VSDs)										
Number of cases	31	9	198	25	7	100	20	27	5	423
Rate (per 1,000 total births)	4.9	2.3	4.4	2.2	2.5	2.8	2.8	4.4	1.2	3.5
Standard Error (SE)	0.88	0.77	0.31	0.44	0.95	0.28	0.62	0.85	0.55	0.17
5. Cleft lip +/- cleft palate										
Number of cases	4	5	51	17	2	44	9	10	3	148
Rate (per 1,000 total births)	0.6	1.3	1.1	1.5	0.7	1.2	1.2	1.6	0.7	1.2
Standard Error (SE)	0.32	0.57	0.16	0.36	0.51	0.18	0.42	0.52	0.43	0.10
6 Hypospadias & epispadias										
Number of cases	11	8	119	15	4	84	16	11	4	272
Rate (per 1,000 total births)	1.8	2.0	2.7	1.3	1.4	2.3	2.2	1.8	1.0	2.2
Standard Error (SE)	0.53	0.72	0.24	0.34	0.72	0.25	0.55	0.54	0.49	0.13
7. Renal dysgenesis or agenesis										
Number of cases	1	1	18	8	2	25	4	3	2	64
Rate (per 1,000 total births)	0.2	0.3	0.4	0.7	0.7	0.7	0.6	0.5	0.5	0.5
Standard Error (SE)	0.16	0.26	0.09	0.25	0.51	0.14	0.28	0.28	0.35	0.07
8. Cystic kidney disease										
Number of cases	5	5	34	9	4	30	7	6	8	108
Rate (per 1,000 total births)	0.8	1.3	0.8	0.8	1.4	0.8	1.0	1.0	2.0	0.9
Standard Error (SE)	0.36	0.57	0.13	0.26	0.72	0.15	0.37	0.40	0.70	0.08
9. Gastroschisis										
Number of cases	2	3	13	5	4	12	6	4	2	51
Rate (per 1,000 total births)	0.3	0.8	0.3	0.4	1.4	0.3	0.8	0.7	0.5	0.4
Standard Error (SE)	0.22	0.44	0.08	0.20	0.72	0.10	0.34	0.33	0.35	0.06
10. Down syndrome										
Number of cases	13	3	130	14	3	92	15	7	8	285
Rate (per 1,000 total births)	2.1	0.8	2.9	1.2	1.1	2.6	2.1	1.1	2.0	2.3
Standard Error (SE)	0.57	0.44	0.25	0.33	0.62	0.27	0.54	0.43	0.70	0.14

Source: 1. Vital Statistics, Birth File, Department of Government Services, March 2006 release.

2. Alberta Congenital Anomalies Surveillance System (ACASS), July 2006 release.

Notes: Data include Alberta residents only.

Totals for Alberta include unknown RHAs.

Data may differ from previously published data due to differences in definitions and dates of data extraction.

4.5 Deaths from Congenital Anomalies

4.5.1 Background

Congenital anomalies are a leading cause of fetal/infant death. Over 1/3 of neonatal deaths and infant mortality are due to congenital anomalies in Alberta. In 1983-2003, there were 6052 infant deaths in Alberta of which 30% to 40% were due to congenital anomalies.

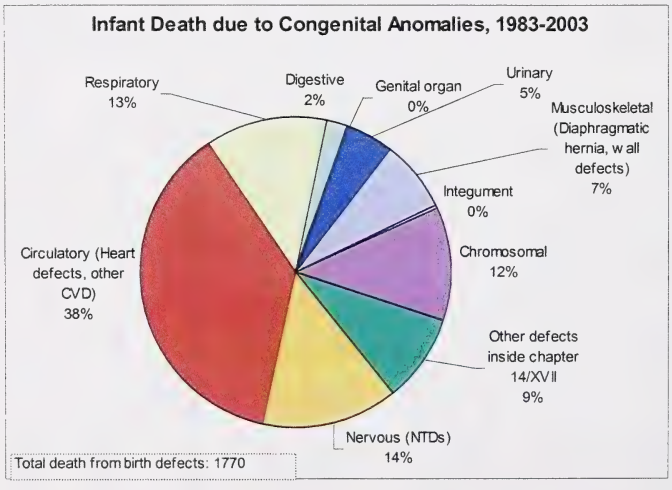
The top five causes of death from congenital anomalies in Alberta are (**Fig. 4.5.1**): circulatory system (heart defects, other defects, 37%), nervous system (NTDs, hydrocephalus, microcephaly, and other defects, 14%), respiratory system (12.8%), chromosomal anomalies (Down syndrome and other chromosome defects, 11.7%), and musculoskeletal (diaphragmatic hernia, gastroschisis, omphalocele, and other musculoskeletal defects, 7.3%).

Over the last two decades, infant deaths from congenital anomalies have decreased significantly for several anomalies in Alberta, e.g., anomalies of the circulatory (i.e., heart defects), nervous (i.e., NTDs), and respiratory systems.

Terminations of affected pregnancies, the decreased incidence of NTDs due to food fortification and wider use of folic acid among pregnant women may have contributed to the reduction in the number of deaths due to congenital anomalies. Improvement in medical procedures for heart defects and other birth defects in recent years may have also contributed to the reduction.

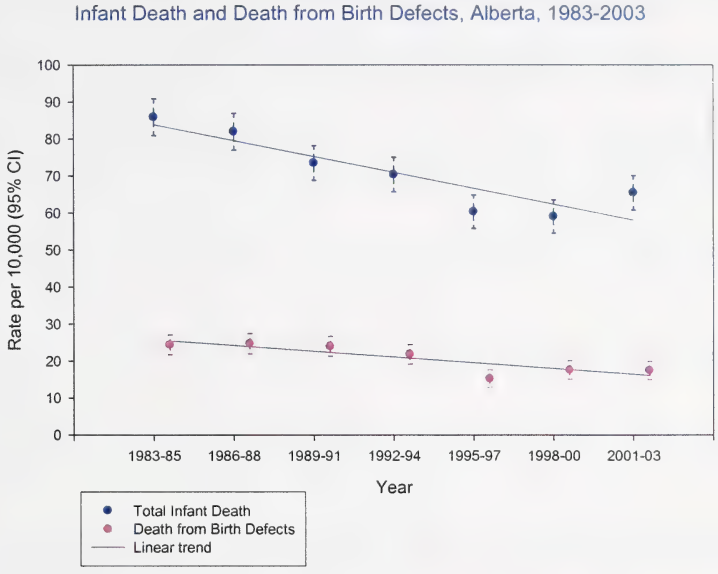
4.5.2 Major Causes of Death by Category

Fig. 4.5.1



4.5.3 Time Trends (see Table 4.5.1)

Fig. 4.5.2



Alberta's death rate (per 10,000 live births) due to congenital anomalies decreased from 24.3 in 1983-85 to 17.4 in 2001-2003. This decrease has contributed in part to the reduction in infant mortality during this period (**Fig. 4.5.2**).

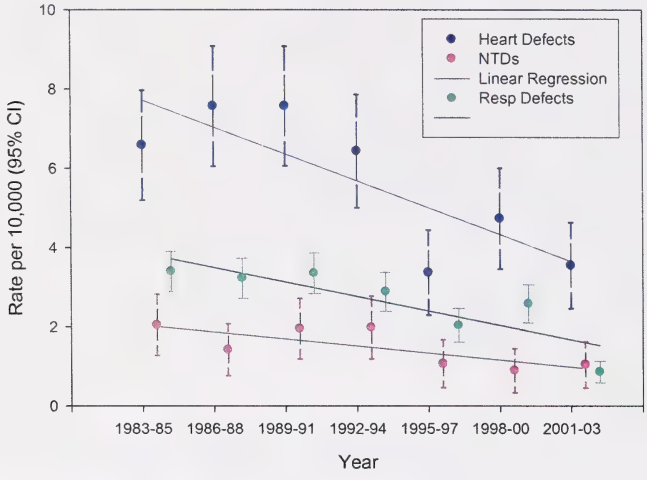
Heart defects, NTDs and respiratory anomalies are three major groups with significant rate decreases from 1983 to 2003 (**Fig. 4.5.3**).

In 2000-03, 201 babies died who had congenital anomalies. Of these, 46 (22.9%) died due to circulatory system anomalies, 29 (14.4%) to nervous system anomalies, and 29 (14.4%) to chromosomal anomalies (five of whom had Down syndrome).

Detailed data on death rates from selected congenital anomalies are presented in **Table 4.5.1**.

Fig. 4.5.3

Infant Death from Selected Congenital Anomalies, Alberta, 1983-2003



4.5.4 Regional Data (see Table 4.5.2)

- The death rate from congenital anomalies was significantly higher than the provincial average in RHA 5 for 2001 to 2003 with a rate of 43.6 (per 10,000 live births; 12 cases). Note the relatively small number of live births compared to other RHAs and also the high standard error (SE) in this region.

- The data from other years by health region are presented in **Table 4.5.2**. There does not appear to be a consistent regional pattern of infant deaths from congenital anomalies over the time period.

Table 4.5.1 Number and Rate of Infant Death from Selected Congenital Anomalies by 3-Year Period, Alberta, 1983 to 2003

Selected anomalies	1983-85	1986-88	1989-91	1992-94	1995-97	1998-00	2001-03	1983-03
1. All congenital anomalies								
Number of deaths	321	312	306	263	171	196	201	1,770
Rate (per 10,000 live births)	24.3	24.6	23.9	21.7	15.2	17.5	17.4	20.9
Standard Error (SE)	1.36	1.39	1.37	1.34	1.16	1.25	1.23	0.50
2. Circulatory system anomalies								
Number of deaths	119	146	121	109	56	63	46	660
Rate (per 10,000 live births)	9.0	11.5	9.5	9.0	5.0	5.6	4.0	7.8
Standard Error (SE)	0.83	0.95	0.86	0.86	0.66	0.71	0.59	0.30
2.1. Heart defects (septal and valve)								
Number of deaths	87	96	97	78	38	53	41	490
Rate (per 10,000 live births)	6.6	7.6	7.6	6.4	3.4	4.7	3.6	5.8
Standard Error (SE)	0.71	0.77	0.77	0.73	0.55	0.65	0.55	0.26
3. Nervous system anomalies								
Number of deaths	52	34	41	41	31	21	29	249
Rate (per 10,000 live births)	3.9	2.7	3.2	3.4	2.8	1.9	2.5	2.9
Standard Error (SE)	0.55	0.46	0.50	0.53	0.49	0.41	0.47	0.19
3.1 Neural tube defects (NTDs)								
Number of deaths	27	18	25	24	12	10	12	128
Rate (per 10,000 live births)	2.0	1.4	2.0	2.0	1.1	0.9	1.0	1.5
Standard Error (SE)	0.39	0.33	0.39	0.40	0.31	0.28	0.30	0.13
4. Respiratory system anomalies								
Number of deaths	45	41	43	35	23	29	10	226
Rate (per 10,000 live births)	3.4	3.2	3.4	2.9	2.0	2.6	0.9	2.7
Standard Error (SE)	0.51	0.51	0.51	0.49	0.43	0.48	0.27	0.18
5. Chromosomal anomalies								
Number of deaths	27	24	33	31	31	32	29	207
Rate (per 10,000 live births)	2.0	1.9	2.6	2.6	2.8	2.9	2.5	2.4
Standard Error (SE)	0.39	0.39	0.45	0.46	0.49	0.51	0.47	0.17
5.1 Non-Down syndrome anomalies (a subset of chromosomal anomalies)								
Number of deaths	22	22	23	27	28	28	24	174
Rate (per 10,000 live births)	1.7	1.7	1.8	2.2	2.5	2.5	2.1	2.1
Standard Error (SE)	0.36	0.37	0.37	0.43	0.47	0.47	0.42	0.16
6. Musculoskeletal system anomalies								
Number of deaths	31	15	20	11	5	23	24	129
Rate (per 10,000 live births)	2.3	1.2	1.6	0.9	0.4	2.1	2.1	1.5
Standard Error (SE)	0.42	0.31	0.35	0.27	0.20	0.43	0.42	0.13
6.1 Diaphragmatic hernia and gastroschisis & omphalocele								
Number of deaths	26	10	11	5	3	13	18	86
Rate (per 10,000 live births)	2.0	0.8	0.9	0.4	0.3	1.2	1.6	1.0
Standard Error (SE)	0.39	0.25	0.26	0.18	0.15	0.32	0.37	0.11
7. Urinary system anomalies								
Number of deaths	13	18	16	10	8	9	16	90
Rate (per 10,000 live births)	1.0	1.4	1.3	0.8	0.7	0.8	1.4	1.1
Standard Error (SE)	0.27	0.33	0.31	0.26	0.25	0.27	0.35	0.11
7.1 Renal dysgenesis or agenesis								
Number of deaths	11	11	15	7	7	4	9	64
Rate (per 10,000 live births)	0.8	0.9	1.2	0.6	0.6	0.4	0.8	0.8
Standard Error (SE)	0.25	0.26	0.30	0.22	0.24	0.18	0.26	0.09

Source: 1. Vital Statistics, Birth File, Department of Government Services, March 2006 release.
2. Vital Statistics, Death File, Department of Government Services, March 2006 release.

Notes: Data include Alberta residents only.
Totals for Alberta include unknown RHAs.
Data may differ from previously published data due to differences in definitions and dates of data extraction.

Table 4.5.2 Number and Rate of Infant Death from Congenital Anomalies by 3-Year Period and Residence RHA, Alberta, 1983-2003

Period of death	R1- Chinook	R2 - Palliser	R3 - Calgary	R4 - David Thompson	R5 - East Central	R6 - Capital	R7 - Aspen	R8 - Peace Country	R9 - Northern Lights	Alberta
1983-85										
Number of deaths	15	5	85	37	12	121	27	15	4	321
Rate (per 10,000 live births)	20.3	12.0	21.0	29.3	26.8	28.2	29.3	22.1	10.1	24.3
Standard Error (SE)	5.24	5.35	2.28	4.80	7.74	2.56	5.63	5.71	5.05	1.36
1986-88										
Number of deaths	8	12	78	30	13	127	19	16	9	312
Rate (per 10,000 live births)	11.7	31.6	19.5	24.1	32.7	30.8	22.0	25.7	25.5	24.6
Standard Error (SE)	4.12	9.10	2.21	4.39	9.05	2.73	5.04	6.42	8.50	1.39
1989-91										
Number of deaths	24	15	87	26	14	94	21	19	6	306
Rate (per 10,000 live births)	35.0	39.5	21.0	21.4	37.3	22.5	24.3	31.7	16.8	23.9
Standard Error (SE)	7.13	10.18	2.24	4.19	9.95	2.32	5.30	7.27	6.87	1.37
1992-94										
Number of deaths	16	10	98	22	6	82	15	8	6	263
Rate (per 10,000 live births)	24.1	28.6	24.9	18.8	17.6	21.1	17.7	13.9	17.8	21.7
Standard Error (SE)	6.02	9.02	2.51	4.00	7.18	2.33	4.58	4.92	7.25	1.34
1995-97										
Number of deaths	12	6	53	18	6	50	10	11	5	171
Rate (per 10,000 live births)	18.9	16.5	14.1	16.5	19.8	14.6	12.8	19.1	14.9	15.2
Standard Error (SE)	5.45	6.73	1.94	3.89	8.08	2.07	4.06	5.75	6.68	1.16
1998-00										
Number of deaths	9	5	62	17	3	59	13	10	8	196
Rate (per 10,000 live births)	14.9	13.5	15.9	15.9	10.5	17.7	17.5	17.6	24.3	17.5
Standard Error (SE)	4.97	6.05	2.02	3.85	6.04	2.30	4.86	5.56	8.59	1.25
2001-03										
Number of deaths	7	6	72	25	12	49	11	13	6	201
Rate (per 10,000 live births)	11.6	16.2	17.5	23.1	43.6	14.4	15.3	22.2	15.9	17.4
Standard Error (SE)	4.39	6.59	2.06	4.61	12.57	2.05	4.60	6.14	6.47	1.23
Total, 1983-2003										
Number of deaths	91	59	535	175	66	582	116	92	44	1,770
Rate (per 10,000 live births)	19.7	22.4	19.2	21.5	27.2	21.9	20.2	21.9	17.7	20.9
Standard Error (SE)	2.07	2.92	0.83	1.62	3.34	0.91	1.88	2.28	2.67	0.50

Source: 1. Vital Statistics, Birth File, Department of Government Services, March 2006 release.
2. Vital Statistics, Death File, Department of Government Services, March 2006 release.

Notes: Data include Alberta residents only.
Totals for Alberta include unknown RHAs.
Data may differ from previously published data due to differences in definitions and dates of data extraction.

4.5.5 Limitations and Methodology Notes

- The causes of death are coded by the International Classification of Diseases (ICD). The 10th revision (ICD-10) replaced the 9th revision in the Vital Statistics Death Registry in 2000. The decrease in death rate for all congenital anomalies may be partly due to changes in classification, rules of underlying cause selection and modification, and coding in ICD-10 (AHW, 2006).
- The ICD codes for death from congenital anomalies are 740-759 for ICD-9 and Q00-Q99 for ICD-10
- Congenital anomalies were grouped by two levels: major sections (by organ systems) and detailed groupings of each section. Groups with large number of deaths were selected to ensure stable rate estimation.
- Coding and reporting practice of causes of death may not be consistent across regions. These differences could contribute to variations in rates across regions and over time

5. SURVEILLANCE AND RESEARCH PROJECTS

In addition to carrying out regular surveillance activities, ACASS also responds to ad hoc requests and conduct research in collaboration with other jurisdictions. Following is an example of these activities.

5.1 Collaboration with International Clearing House of Birth Defects

5.1.1 Neural Tube Defects (NTDs) and Anal defects

Requests for data/information
2005
NTDs and maternal age – Alberta Health, Provincial Health Office
NTD rates – Calgary genetics
ICD-10 coding system – Alberta Health
Cleft lip and palate – International Clearinghouse of Birth Defects Surveillance and Research (ICBDSR)
Sirenomelia – Calgary genetics
Gastroschisis x2 – ICBDSR and Alberta Regional Health Authority (RHA)
CNS anomalies – Calgary genetics
Congenital heart defects – University of Calgary (U of C) researcher
Limb reductions – Alberta RHA
Congenital anomalies and neonatal deaths – Alberta Perinatal Health Programme (APHP)
Factors to consider when establishing a surveillance system – University of Saskatchewan
Congenital anomaly rates – Calgary Health Region
Pierre Robin – media
Congenital anomaly data – B.C. MSc student

2004
Congenital Heart Disease prevalence – Calgary cardiologist
Down Syndrome – Calgary genetics
Congenital Heart Defects in endogamous population – Calgary genetics
Ascertainment methods/sources – Alberta Health, Provincial Health Office
Birth defect rates – Calgary genetics
Albinism – Calgary genetics
Diaphragmatic hernia in an endogamous population – Calgary genetics
Holoprosencephaly – Calgary neonatologist
NTDs in an endogamous population – Calgary genetics
Cleft lip and palate – Calgary genetics

2003
NTDs – Alberta RHA
Anophthalmia/microphthalmia – Calgary genetics
Information re establishing a surveillance system – Saskatoon
Information re establishing surveillance system – Saskatchewan Health Department
Small intestinal atresia – Calgary genetics
ACASS data comparison with North Central Alberta Perinatal database – Health Surveillance
NTDs, Cleft lip and palate, Down Syndrome – Calgary genetics

Cystic Fibrosis – Research assistant, Calgary
2002
Limb reductions – Calgary genetics
Limb reductions – Canadian Congenital Anomalies Surveillance System (CCASS)
Birth defect rates – media
Bowen-Conradi – Calgary genetics
Cloacal Exstrophy – Saskatoon genetics
Coding issues – CCASS
Pyloric stenosis – Calgary genetics
Down Syndrome – Alberta Children's Hospital (ACH)
Down Syndrome – parent support group

2001
Congenital anomalies with vascular pathogenesis – Ottawa and Calgary genetics
Single umbilical artery – community genetics
NTDs – Calgary research assistant
Down Syndrome – ACH
Choanal atresia – CCASS
Info re establishing a surveillance system – Hospital for Sick Children, Toronto
Diaphragmatic hernia – researcher
Cystic Fibrosis (CF) rates – Calgary genetics
Thalassemia rates – Calgary genetics

5.2 Surveillance and Research Projects/Collaborations and Consultations/Papers

Consultation:
Calgary Health Region – Healthy Communities: re promotion of folic acid in community and in school curricula

Papers:

1. De Wals P et al. Impact of folic acid food fortification on the prevalence of neural tube defects in Canada (submitted for publication)
2. Lowry RB, Sibbald B and Bedard T. Stability of prevalence rates of anorectal malformations in the Alberta Congenital Anomalies Surveillance System 1990-2004. Journal of Pediatric Surgery (in press).
3. Paquette D, Lowry RB and Sauvé R. Two to three percent of infants are born with a congenital anomaly, but who's counting? A national survey of congenital anomalies surveillance in Canada. Chronic Dis Can. 2006; 27(1): 3-5.
4. Lowry RB, Kohut R, Sibbald B & Rouleau J. Anophthalmia and microphthalmia in the Alberta Congenital Anomalies Surveillance System. Can J Ophthalmol 2005; 40: 38-44
5. Lowry RB, Sibbald B, Bamforth JS Re: An epidemiologic analysis of CHARGE Syndrome: preliminary results from a Canadian study (letter). Am J Med Gen 2005; 139A: 169
6. Wang FL, Gabos S, Sibbald B, Lowry RB Completeness and accuracy of the birth registry data on congenital anomalies in Alberta, Canada Chronic Diseases in Canada 2001; 22(2): 57-66
7. Articles for Canadian Congenital Anomalies Surveillance System Current Contents (http://www.phac-aspc.gc.ca/ccasn-rcsac/index.html):
    i. Sibbald B and Lowry RB Orofacial clefts in Alberta 1980-2004 inclusive (winter 2005) http://www.phac-aspc.gc.ca/ccasn-rcsac/ct2005/or-cl-alberta_e.html
    ii. Sibbald B and Lowry RB Abdominal wall defects- Alberta 1980-2002 (winter 2004) http://www.phac-aspc.gc.ca/ccasn-rcsac/ct2004/awd-alb.html
    iii. Sibbald B and Lowry RB Down Syndrome in Alberta: Alberta Congenital Anomalies Surveillance System (fall 2003) http://www.phac-aspc.gc.ca/ccasn-rcsac/ct2003/abds_e.html

Alberta Congenital anomalies Surveillance System

6. APPENDICES

Appendix A1 Flow Chart of the Process of ACASS Data Collection

Appendix A2 Congenital Anomaly(ies) Reporting Form (CARF)

Appendix A3 Anomaly rates per 1,000 total births

Appendix A4 Selected anomalies with rates of live births and stillbirths compared with total rates including terminations of pregnancy (ToP)

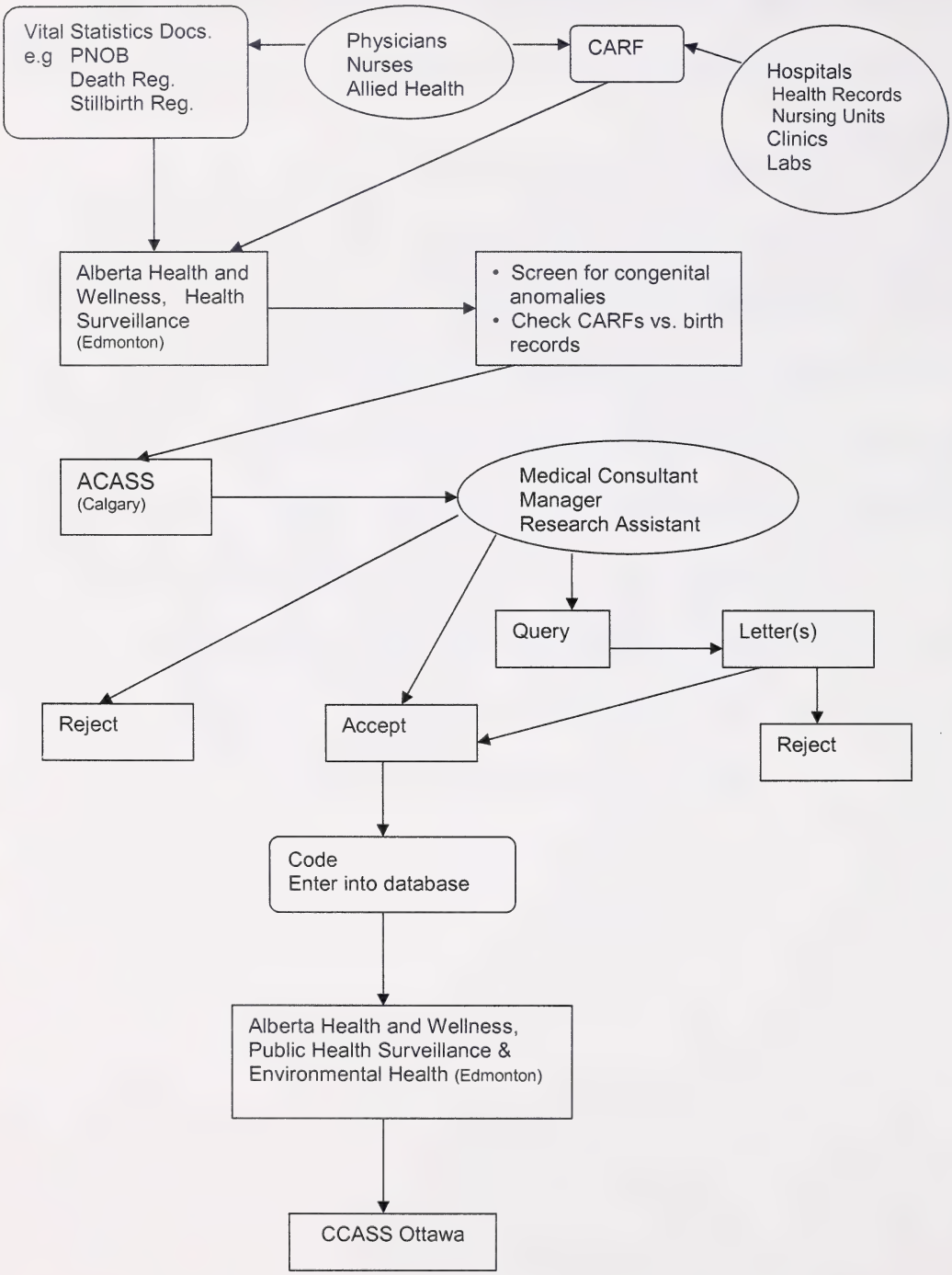
Appendix A5 Numbers of cases, anomalies and anomalies per case 1980-2004

Appendix A6 Termination of pregnancy (ToP) for Congenital anomalies

Appendix A.7 Diagram of Embryonic and Fetal Developmental Stage and Diagnosis of Congenital Anomalies

Appendix A.8 Critical Periods of Embryogenesis by Major Organs/Systems in Humans

Appendix A.1 Flow Chart of the Process of ACASS Data Collection



Alberta

Appendix A.2 Congenital Anomaly(ies) Reporting Form (CARF)

Alberta
HEALTH AND WELLNESS

Congenital Anomaly(ies) Reporting Form

PLEASE TYPE OR PRINT CLEARLY

Fetus / Infant:

Name: (Last, First, Initial)

Gender: ☐ Male ☐ Female ☐ Unknown

Type of Birth: ☐ Livebirth ☐ Stillbirth ☐ Fetus less than 20 weeks gestation

Name of Hospital of Birth:

Birthweight: Grams

Gestation Age: (Completed Weeks)

Location of Hospital of Birth: (City/Town)

Child's personal Health Number:

Attending Physician's Name:

Plurality of Birth: ☐ Single ☐ Twin ☐ First ☐ Second ☐ Triplets ☐ First ☐ Second ☐ Third

Physician Responsible for Ongoing Care: (if different from above)

Parents:

Mother's Name: (Last, First, Maiden)

Date of Birth or Age (if DOB unknown)

Month by Name Day Year

Permanent Address:

Mother's Personal Health Number:

City/Town: Postal Code:

Father's Name: (Last, First, Initial)

Date of Birth or Age (if DOB unknown)

Month by Name Day Year

Total Number

Livebirth

Stillbirths

Spontaneous Abortions

Therapeutic Abortions

Reporting Hospital/Agency/Clinic

Name:

Infant's Admission (if different from birthdate)

Month by Name Day Year

Infant's Discharge

Month by Name Day Year

Location: (City/Town)

Infant's Death (if applicable)

Month by Name Day Year

Full description of Congenital Anomaly(ies) and/or **SYNDROME DIAGNOSES**
(if necessary, please attach supporting documents)

OFFICE USE ONLY

Completed by: Position: Date:

HS 320-112 (98/12) Alberta Congenital Anomalies Surveillance Reporting Hospital Division of Vital Statistics

Alberta

Appendix A.3 Alberta Congenital Anomalies Surveillance System, ICD-9 Chapter XIV/ICD-10 Chapter XVII Anomaly Rates per 1,000 Total Births (Live Births + Stillbirths)

Diagnostic Category		80-89	90-99	80-99	00-04
Anencephaly	NUMBER	147	75	222	26
	RATE	**0.34**	**0.19**	**0.27**	**0.13**
	Lower CI	0.29	0.15	0.23	0.09
	Upper CI	0.40	0.24	0.31	0.20
Spina Bifida without Anencephaly	NUMBER	218	190	408	49
	RATE	**0.51**	**0.48**	**0.49**	**0.25**
	Lower CI	0.44	0.41	0.45	0.19
	Upper CI	0.58	0.55	0.54	0.33
Encephalocele	NUMBER	43	34	77	24
	RATE	**0.10**	**0.09**	**0.09**	**0.12**
	Lower C	0.07	0.06	0.07	0.08
	Upper CI	0.13	0.12	0.12	0.18
Neural Tube Defects (all)	NUMBER	408	299	707	99
	RATE	**0.95**	**0.75**	**0.85**	**0.51**
	Lower C	0.86	0.67	0.79	0.42
	Upper CI	1.04	0.84	0.92	0.62
Hydrocephalus without Spina Bifida	NUMBER	237	193	430	114
	RATE	**0.55**	**0.49**	**0.52**	**0.59**
	Lower C	0.48	0.42	0.47	0.49
	Upper CI	0.63	0.56	0.57	0.71
Microcephaly	NUMBER	157	117	274	69
	RATE	**0.36**	**0.30**	**0.33**	**0.36**
	Lower C	0.31	0.24	0.29	0.28
	Upper CI	0.43	0.35	0.37	0.45
Anophthalmia/microphthalmia	NUMBER	55	62	117	28
	RATE	**0.13**	**0.16**	**0.14**	**0.14**
	Lower C	0.10	0.12	0.12	0.10
	Upper CI	0.17	0.20	0.17	0.21
Congenital cataract	NUMBER	60	54	114	16
	RATE	**0.14**	**0.14**	**0.14**	**0.08**
	Lower C	0.11	0.10	0.11	0.05
	Upper CI	0.18	0.18	0.17	0.13
Aniridia	NUMBER	1	4	5	3
	RATE	**0.00**	**0.01**	**0.01**	**0.02**
	Lower C	0.00	0.00	0.00	0.00
	Upper CI	0.01	0.03	0.01	0.04
Anotia/microtia	NUMBER	27	57	84	39
	RATE	**0.06**	**0.14**	**0.10**	**0.20**
	Lower C	0.04	0.11	0.08	0.14
	Upper CI	0.09	0.19	0.13	0.28
Common Truncus	NUMBER	38	30	68	9
	RATE	**0.09**	**0.08**	**0.08**	**0.05**
	Lower C	0.06	0.05	0.06	0.02
	Upper CI	0.12	0.11	0.10	0.09

Appendix A.3 Alberta Congenital Anomalies Surveillance System, ICD-9 Chapter XIV/ICD-10 Chapter XVII Anomaly Rates per 1,000 Total Births (Live Births + Stillbirths)

Diagnostic Category		2000	2001	2002	2003	2004	2005
Anencephaly	NUMBER	8	3	3	9	3	7
	RATE	**0.22**	**0.08**	**0.08**	**0.22**	**0.07**	**0.17**
	Lower CI	0.09	0.02	0.02	0.10	0.01	0.07
	Upper CI	0.42	0.23	0.22	0.42	0.21	0.34
Spina Bifida without Anencephaly	NUMBER	7	12	6	11	13	8
	RATE	**0.19**	**0.32**	**0.16**	**0.27**	**0.32**	**0.19**
	Lower CI	0.08	0.17	0.06	0.14	0.17	0.08
	Upper CI	0.39	0.56	0.33	0.49	0.55	0.37
Encephalocele	NUMBER	5	7	6	3	3	6
	RATE	**0.14**	**0.19**	**0.16**	**0.07**	**0.07**	**0.14**
	Lower CI	0.04	0.08	0.06	0.02	0.01	0.05
	Upper CI	0.31	0.38	0.33	0.21	0.21	0.31
Neural Tube Defects (all)	NUMBER	20	22	15	23	19	21
	RATE	**0.54**	**0.59**	**0.39**	**0.57**	**0.47**	**0.50**
	Lower CI	0.33	0.37	0.22	0.36	0.28	0.31
	Upper CI	0.84	0.89	0.64	0.86	0.73	0.77
Hydrocephalus without Spina Bifida	NUMBER	28	21	21	22	22	18
	RATE	**0.76**	**0.56**	**0.55**	**0.55**	**0.54**	**0.43**
	Lower CI	0.51	0.35	0.34	0.34	0.34	0.26
	Upper CI	1.10	0.86	0.83	0.83	0.82	0.68
Microcephaly	NUMBER	13	19	8	19	10	16
	RATE	**0.35**	**0.51**	**0.21**	**0.47**	**0.25**	**0.38**
	Lower CI	0.19	0.31	0.09	0.29	0.12	0.22
	Upper CI	0.60	0.79	0.41	0.74	0.45	0.62
Anophthalmia/microphthalmia	NUMBER	12	5	2	1	8	10
	RATE	**0.33**	**0.13**	**0.05**	**0.02**	**0.20**	**0.24**
	Lower CI	0.17	0.04	0.01	0.00	0.09	0.11
	Upper CI	0.57	0.31	0.18	0.13	0.39	0.44
Congenital cataract	NUMBER	3	3	2	4	4	4
	RATE	**0.08**	**0.08**	**0.05**	**0.10**	**0.10**	**0.10**
	Lower CI	0.02	0.02	0.01	0.03	0.03	0.03
	Upper CI	0.23	0.23	0.18	0.25	0.25	0.24
Aniridia	NUMBER	0	0	0	2	1	0
	RATE				**0.05**	**0.02**	
	Lower CI				0.01	0.00	
	Upper CI				0.17	0.12	
Anotia/microtia	NUMBER	7	6	6	7	13	11
	RATE	**0.19**	**0.16**	**0.16**	**0.17**	**0.32**	**0.26**
	Lower CI	0.06	0.06	0.06	0.07	0.17	0.13
	Upper CI	0.34	0.34	0.33	0.36	0.55	0.47
Common Truncus	NUMBER	3	1	2	2	1	2
	RATE	**0.08**	**0.03**	**0.05**	**0.05**	**0.02**	**0.05**
	Lower CI	0.02	0.00	0.01	0.01	0.02	0.01
	Upper CI	0.23	0.14	0.18	0.17	0.12	0.16

Appendix A.3 Alberta Congenital Anomalies Surveillance System, ICD-9 Chapter XIV/ICD-10 Chapter XVII Anomaly Rates per 1,000 Total Births (Live Births + Stillbirths)

Diagnostic Category		80-89	90-99	80-99	00-04
Transposition of Great Arteries	NUMBER	129	128	257	93
	RATE	**0.30**	**0.32**	**0.31**	**0.48**
	Lower CI	0.25	0.27	0.27	0.39
	Upper CI	0.36	0.38	0.35	0.59
Tetralogy of Fallot	NUMBER	99	115	214	56
	RATE	**0.23**	**0.29**	**0.26**	**0.29**
	Lower CI	0.19	0.24	0.23	0.22
	Upper CI	0.28	0.35	0.30	0.38
Ventricular Septal Defect	NUMBER	1198	1128	2326	600
	RATE	**2.78**	**2.84**	**2.81**	**3.10**
	Lower CI	2.63	2.68	2.70	2.86
	Upper CI	2.94	3.02	2.93	3.36
Atrial Septal Defect	NUMBER	528	762	1290	366
	RATE	**1.23**	**1.92**	**1.56**	**1.89**
	Lower CI	1.12	1.79	1.48	1.70
	Upper CI	1.34	2.06	1.65	2.10
Endocardial Cushion Defect	NUMBER	147	152	299	90
	RATE	**0.34**	**0.38**	**0.36**	**0.47**
	Lower CI	0.29	0.32	0.32	0.37
	Upper CI	0.40	0.45	0.40	0.57
Pulmonary Valve Atresia and Stenosis	NUMBER	333	249	582	108
	RATE	**0.77**	**0.63**	**0.70**	**0.56**
	Lower CI	0.69	0.55	0.65	0.46
	Upper CI	0.86	0.71	0.76	0.67
Tricuspid Valve Atresia and Stenosis	NUMBER	41	33	74	12
	RATE	**0.10**	**0.08**	**0.09**	**0.06**
	Lower CI	0.07	0.06	0.07	0.03
	Upper CI	0.13	0.12	0.11	0.11
Ebstein's Anomaly	NUMBER	20	20	40	12
	RATE	**0.05**	**0.05**	**0.05**	**0.06**
	Lower CI	0.03	0.03	0.03	0.03
	Upper CI	0.07	0.08	0.07	0.11
Aortic Valve Stenosis	NUMBER	61	78	139	44
	RATE	**0.14**	**0.20**	**0.17**	**0.23**
	Lower CI	0.11	0.16	0.14	0.17
	Upper CI	0.18	0.25	0.20	0.31
Hypoplastic Left Heart Syndrome	NUMBER	95	87	182	47
	RATE	**0.22**	**0.22**	**0.22**	**0.24**
	Lower CI	0.18	0.18	0.19	0.18
	Upper CI	0.27	0.27	0.25	0.32
Patent Ductus Arteriosus	NUMBER	945	102	1265	128
	RATE	**2.19**	**0.54**	**1.53**	**0.66**
	Lower CI	2.06	0.44	1.45	0.55
	Upper CI	2.34	0.65	1.62	0.79

Appendix A.3 Alberta Congenital Anomalies Surveillance System, ICD-9 Chapter XIV/ICD-10 Chapter XVII Anomaly Rates per 1,000 Total Births (Live Births + Stillbirths)

Diagnostic Category		2000	2001	2002	2003	2004	2005
Transposition of Great Arteries	NUMBER	11	23	21	18	20	17
	RATE	**0.30**	**0.61**	**0.55**	**0.45**	**0.49**	**0.41**
	Lower CI	0.15	0.39	0.34	0.27	0.30	0.24
	Upper CI	0.53	0.92	0.83	0.71	0.76	0.65
Tetralogy of Fallot	NUMBER	14	9	11	10	12	9
	RATE	**0.38**	**0.24**	**0.29**	**0.25**	**0.30**	**0.22**
	Lower CI	0.21	0.11	0.14	0.12	0.15	0.10
	Upper CI	0.64	0.45	0.51	0.46	0.52	0.41
Ventricular Septal Defect	NUMBER	99	102	135	111	153	131
	RATE	**2.69**	**2.72**	**3.50**	**2.77**	**3.77**	**3.13**
	Lower CI	2.19	2.22	2.94	2.28	3.20	2.62
	Upper CI	3.27	3.31	4.15	3.33	4.42	3.72
Atrial Septal Defect	NUMBER	53	57	74	86	96	76
	RATE	**1.44**	**1.52**	**1.92**	**2.14**	**2.37**	**1.82**
	Lower CI	1.08	1.15	1.51	1.72	1.92	1.43
	Upper CI	1.88	1.97	2.41	2.65	2.89	2.27
Endocardial Cushion Defect	NUMBER	16	24	11	19	20	13
	RATE	**0.43**	**0.64**	**0.29**	**0.47**	**0.49**	**0.31**
	Lower CI	0.25	0.41	0.14	0.29	0.30	0.17
	Upper CI	0.70	0.95	0.51	0.74	0.76	0.53
Pulmonary Valve Atresia and Stenosis	NUMBER	18	26	23	23	18	25
	RATE	**0.49**	**0.69**	**0.60**	**0.57**	**0.44**	**0.60**
	Lower CI	0.29	0.45	0.38	0.36	0.26	0.39
	Upper CI	0.77	1.02	0.90	0.86	0.70	0.88
Tricuspid Valve Atresia and Stenosis	NUMBER	1	3	1	4	3	1
	RATE	**0.03**	**0.08**	**0.03**	**0.10**	**0.01**	**0.02**
	Lower CI	0.00	0.02	0.00	0.03	0.01	0.00
	Upper CI	0.14	0.23	0.13	0.25	0.21	0.12
Ebstein's Anomaly	NUMBER	3	2	4	2	1	1
	RATE	**0.08**	**0.05**	**0.10**	**0.05**	**0.02**	**0.02**
	Lower CI	0.02	0.01	0.03	0.01	0.00	0.00
	Upper CI	0.23	0.18	0.26	0.17	0.12	0.12
Aortic Valve Stenosis	NUMBER	10	6	9	8	11	3
	RATE	**0.27**	**0.16**	**0.23**	**0.20**	**0.27**	**0.07**
	Lower CI	0.13	0.06	0.11	0.09	0.14	0.01
	Upper CI	0.50	0.34	0.44	0.39	0.48	0.20
Hypoplastic Left Heart Syndrome	NUMBER	15	12	6	6	8	18
	RATE	**0.41**	**0.32**	**0.16**	**0.15**	**0.20**	**0.43**
	Lower CI	0.23	0.17	0.06	0.05	0.09	0.26
	Upper CI	0.67	0.56	0.33	0.32	0.39	0.68
Patent Ductus Arteriosus	NUMBER	24	18	23	29	34	31
	RATE	**0.65**	**0.48**	**0.60**	**0.72**	**0.84**	**0.74**
	Lower CI	0.42	0.29	0.38	0.48	0.58	0.50
	Upper CI	0.97	0.76	0.90	1.04	1.17	1.05

Appendix A.3 Alberta Congenital Anomalies Surveillance System, ICD-9 Chapter XIV/ICD-10 Chapter XVII Anomaly Rates per 1,000 Total Births (Live Births + Stillbirths)

Diagnostic Category		80-89	90-99	80-99	00-04
Coarctation of the Aorta	NUMBER	173	196	369	76
	RATE	**0.40**	**0.49**	**0.45**	**0.39**
	Lower CI	0.34	0.43	0.40	0.31
	Upper CI	0.47	0.57	0.49	0.49
Cleft Palate without Cleft Lip	NUMBER	281	331	612	157
	RATE	**0.65**	**0.83**	**0.74**	**0.81**
	Lower CI	0.58	0.75	0.68	0.69
	Upper CI	0.73	0.93	0.80	0.95
Cleft Lip with and without Cleft Palate	NUMBER	478	461	939	228
	RATE	**1.11**	**1.16**	**1.14**	**1.18**
	Lower CI	1.01	1.06	1.06	1.03
	Upper CI	1.21	1.27	1.21	1.34
Choanal Atresia	NUMBER	51	40	91	39
	RATE	**0.12**	**0.10**	**0.11**	**0.20**
	Lower CI	0.09	0.07	0.09	0.14
	Upper CI	0.16	0.14	0.14	0.28
Oesophageal Atresia/ Tracheo-oesphageal Fistula	NUMBER	128	93	221	36
	RATE	**0.30**	**0.23**	**0.27**	**0.19**
	Lower CI	0.25	0.19	0.23	0.13
	Upper CI	0.35	0.29	0.30	0.26
Rectal and Large Intestinal Atresia/Stenosis	NUMBER	202	212	414	124
	RATE	**0.47**	**0.53**	**0.50**	**0.64**
	Lower CI	0.41	0.47	0.45	0.53
	Upper CI	0.54	0.61	0.55	0.76
Pyloric Stenosis	NUMBER	386	281	667	179
	RATE	**0.90**	**0.71**	**0.81**	**0.93**
	Lower CI	0.81	0.63	0.75	0.79
	Upper CI	0.99	0.80	0.87	1.07
Hirschsprung Disease	NUMBER	63	55	118	23
	RATE	**0.15**	**0.14**	**0.14**	**0.12**
	Lower CI	0.11	0.10	0.12	0.08
	Upper CI	0.19	0.18	0.17	0.18
Biliary Atresia	NUMBER	20	24	44	12
	RATE	**0.05**	**0.06**	**0.05**	**0.06**
	Lower CI	0.03	0.04	0.04	0.03
	Upper CI	0.07	0.09	0.07	0.11
Renal Agenesis/Hypoplasia	NUMBER	168	187	355	103
	RATE	**0.39**	**0.47**	**0.43**	**0.53**
	Lower CI	0.33	0.41	0.39	0.43
	Upper CI	0.45	0.54	0.48	0.65
Bladder Exstrophy	NUMBER	12	11	23	8
	RATE	**0.03**	**0.03**	**0.03**	**0.04**
	Lower CI	0.01	0.01	0.02	0.02
	Upper CI	0.05	0.05	0.04	0.08

Appendix A.3 Alberta Congenital Anomalies Surveillance System, ICD-9 Chapter XIV/ICD-10 Chapter XVII Anomaly Rates per 1,000 Total Births (Live Births + Stillbirths)

Diagnostic Category		2000	2001	2002	2003	2004	2005
Coarctation of the Aorta	NUMBER	16	13	20	14	13	14
	RATE	**0.43**	**0.35**	**0.52**	**0.35**	**0.32**	**0.33**
	Lower CI	0.25	0.19	0.32	0.19	0.17	0.18
	Upper CI	0.70	0.59	0.80	0.58	0.55	0.56
Cleft Palate without Cleft Lip	NUMBER	31	34	34	24	34	26
	RATE	**0.84**	**0.91**	**0.88**	**0.60**	**0.84**	**0.62**
	Lower CI	0.57	0.63	0.61	0.38	0.58	0.41
	Upper CI	1.19	1.27	1.23	0.89	1.17	0.91
Cleft Lip with and without Cleft Palate	NUMBER	40	45	52	45	46	44
	RATE	**1.09**	**1.20**	**1.35**	**1.12**	**1.13**	**1.05**
	Lower CI	0.78	0.88	1.01	0.82	0.83	0.77
	Upper CI	1.48	1.61	1.77	1.50	1.51	1.41
Choanal Atresia	NUMBER	2	11	7	6	9	7
	RATE	**0.16**	**0.29**	**0.18**	**0.15**	**0.22**	**0.17**
	Lower CI	0.06	0.15	0.07	0.05	0.10	0.07
	Upper CI	0.35	0.52	0.37	0.32	0.42	0.34
Oesophageal Atresia/ Tracheo-oesphageal Fistula	NUMBER	5	6	9	6	10	7
	RATE	**0.14**	**0.16**	**0.23**	**0.15**	**0.25**	**0.17**
	Lower CI	0.04	0.06	0.11	0.05	0.12	0.07
	Upper CI	0.31	0.34	0.44	0.32	0.45	0.34
Rectal and Large Intestinal Atresia/Stenosis	NUMBER	21	26	25	25	27	15
	RATE	**0.57**	**0.69**	**0.65**	**0.62**	**0.67**	**0.36**
	Lower CI	0.35	0.45	0.42	0.40	0.44	0.20
	Upper CI	0.87	1.02	0.96	0.92	0.97	0.59
Pyloric Stenosis	NUMBER	33	36	37	40	33	40
	RATE	**0.90**	**0.96**	**0.96**	**1.00**	**0.81**	**0.96**
	Lower CI	0.62	0.67	0.68	0.71	0.56	0.68
	Upper CI	1.26	1.33	1.32	1.36	1.14	1.30
Hirschsprung Disease	NUMBER	4	3	3	8	5	5
	RATE	**0.11**	**0.08**	**0.08**	**0.20**	**0.12**	**0.12**
	Lower CI	0.03	0.02	0.02	0.09	0.04	0.04
	Upper CI	0.27	0.22	0.22	0.39	0.28	0.27
Biliary Atresia	NUMBER	1	2	2	2	5	4
	RATE	**0.03**	**0.05**	**0.05**	**0.05**	**0.12**	**0.10**
	Lower CI	0.00	0.01	0.01	0.01	0.04	0.03
	Upper CI	0.14	0.18	0.18	0.17	0.28	0.24
Renal Agenesis/Hypoplasia	NUMBER	19	19	25	18	22	15
	RATE	**0.52**	**0.51**	**0.65**	**0.45**	**0.54**	**0.36**
	Lower CI	0.31	0.31	0.42	0.27	0.34	0.20
	Upper CI	0.80	0.79	0.96	0.71	0.82	0.59
Bladder Exstrophy	NUMBER	1	4	1	2	0	1
	RATE	**0.03**	**0.11**	**0.03**	**0.05**		**0.02**
	Lower CI	0.00	0.03	0.00	0.01		0.00
	Upper CI	0.14	0.27	0.13	0.17		0.12

Appendix A.3 Alberta Congenital Anomalies Surveillance System, ICD-9 Chapter XIV/ICD-10 Chapter XVII Anomaly Rates per 1,000 Total Births (Live Births + Stillbirths)

Diagnostic Category		80-89	90-99	80-99	00-04
Obstructive Genitourinary Defect	NUMBER	399	658	1057	446
	RATE	**0.93**	**1.66**	**1.28**	**2.30**
	Lower CI	0.84	1.54	1.20	2.10
	Upper CI	1.02	1.79	1.36	2.53
Hypospadias and Epispadias †	NUMBER	912	852	1764	413
	RATE	**4.16**	**4.19**	**4.17**	**4.17**
	Lower CI	3.89	3.91	3.98	3.78
	Upper CI	4.43	4.48	4.37	4.59
Reduction Deformity, Upper Limbs	NUMBER	224	256	480	121
	RATE	**0.52**	**0.65**	**0.58**	**0.63**
	Lower CI	0.45	0.57	0.53	0.52
	Upper CI	0.59	0.73	0.63	0.75
Reduction Deformity, Lower Limbs	NUMBER	111	124	235	72
	RATE	**0.26**	**0.31**	**0.28**	**0.37**
	Lower CI	0.21	0.26	0.25	0.29
	Upper CI	0.31	0.37	0.32	0.47
Gastroschisis	NUMBER	61	80	141	61
	RATE	**0.14**	**0.20**	**0.17**	**0.32**
	Lower CI	0.11	0.16	0.14	0.24
	Upper CI	0.18	0.25	0.20	0.41
Omphalocele	NUMBER	87	75	162	39
	RATE	**0.20**	**0.19**	**0.20**	**0.20**
	Lower CI	0.16	0.15	0.17	0.14
	Upper CI	0.25	0.24	0.23	0.28
Congenital Hip Dislocation	NUMBER	616	453	1069	129
	RATE	**1.43**	**1.14**	**1.29**	**0.67**
	Lower CI	1.32	1.04	1.22	0.56
	Upper CI	1.55	1.25	1.37	0.79
Diaphragmatic Hernia	NUMBER	142	103	245	68
	RATE	**0.33**	**0.26**	**0.30**	**0.35**
	Lower CI	0.28	0.21	0.26	0.27
	Upper CI	0.39	0.31	0.34	0.45
Trisomy 13	NUMBER	32	47	79	26
	RATE	**0.07**	**0.12**	**0.10**	**0.13**
	Lower CI	0.05	0.09	0.08	0.09
	Upper CI	0.10	0.16	0.12	0.20
Down Syndrome (Trisomy 21)	NUMBER	403	451	854	300
	RATE	**0.94**	**1.14**	**1.03**	**1.55**
	Lower CI	0.85	1.03	0.96	1.38
	Upper CI	1.03	1.25	1.10	1.74
Trisomy 18	NUMBER	74	105	179	57
	RATE	**0.17**	**0.26**	**0.22**	**0.29**
	Lower CI	0.14	0.22	0.19	0.22
	Upper CI	0.22	0.32	0.25	0.38

Appendix A.3 Alberta Congenital Anomalies Surveillance System, ICD-9 Chapter XIV/ICD-10 Chapter XVII Anomaly Rates per 1,000 Total Births (Live Births + Stillbirths)

Diagnostic Category		2000	2001	2002	2003	2004	2005
Obstructive Genitourinary Defect	NUMBER	68	100	87	95	96	88
	RATE	**1.85**	**2.67**	**2.26**	**2.37**	**2.37**	**2.10**
	Lower CI	1.43	2.17	1.81	1.92	1.92	1.69
	Upper CI	2.34	3.25	2.78	2.89	2.89	2.59
Hypospadias and Epispadias†	NUMBER	85	81	64	89	94	97
	RATE	**4.54**	**4.21**	**3.25**	**4.33**	**4.52**	**4.54**
	Lower CI	3.63	3.35	2.50	3.48	3.65	3.68
	Upper CI	5.61	5.23	4.14	5.33	5.53	5.54
Reduction Deformity, Upper Limbs	NUMBER	18	26	24	30	23	20
	RATE	**0.49**	**0.69**	**0.62**	**0.75**	**0.57**	**0.48**
	Lower CI	0.29	0.45	0.40	0.51	0.36	0.29
	Upper CI	0.79	1.02	0.93	1.07	0.85	0.74
Reduction Deformity, Lower Limbs	NUMBER	15	16	17	11	13	12
	RATE	**0.41**	**0.43**	**0.44**	**0.27**	**0.32**	**0.29**
	Lower CI	0.23	0.24	0.26	0.14	0.17	0.15
	Upper CI	0.67	0.69	0.71	0.49	0.55	0.50
Gastroschisis	NUMBER	8	11	16	13	13	22
	RATE	**0.22**	**0.29**	**0.42**	**0.32**	**0.32**	**0.53**
	Lower CI	0.09	0.15	0.24	0.17	0.17	0.33
	Upper CI	0.42	0.52	0.67	0.55	0.55	0.80
Omphalocele	NUMBER	8	7	8	8	8	3
	RATE	**0.22**	**0.19**	**0.21**	**0.20**	**0.20**	**0.07**
	Lower CI	0.09	0.08	0.09	0.09	0.09	0.01
	Upper CI	0.42	0.38	0.41	0.39	0.39	0.20
Congenital Hip Dislocation	NUMBER	38	30	21	15	25	22
	RATE	**1.03**	**0.80**	**0.55**	**0.37**	**0.62**	**0.53**
	Lower CI	0.73	0.54	0.34	0.21	0.40	0.33
	Upper CI	1.42	1.14	0.83	0.62	0.91	0.80
Diaphragmatic Hernia	NUMBER	19	17	16	11	5	15
	RATE	**0.52**	**0.45**	**0.42**	**0.27**	**0.12**	**0.36**
	Lower CI	0.31	0.26	0.24	0.14	0.04	0.20
	Upper CI	0.80	0.73	0.67	0.49	0.28	0.59
Trisomy 13	NUMBER	7	3	4	5	7	4
	RATE	**0.19**	**0.08**	**0.10**	**0.12**	**0.17**	**0.10**
	Lower CI	0.08	0.02	0.03	0.04	0.07	0.03
	Upper CI	0.39	0.23	0.26	0.29	0.35	0.24
Down Syndrome (Trisomy 21)	NUMBER	54	57	50	72	67	84
	RATE	**1.47**	**1.52**	**1.30**	**1.80**	**1.65**	**20.1**
	Lower CI	1.10	1.15	0.96	1.41	1.28	1.60
	Upper CI	1.91	1.97	1.71	2.26	2.10	2.49
Trisomy 18	NUMBER	13	15	12	5	12	18
	RATE	**0.35**	**0.40**	**0.31**	**0.12**	**0.30**	**0.43**
	Lower CI	0.19	0.22	0.16	0.04	0.15	0.26
	Upper CI	0.60	0.66	0.54	0.29	0.52	0.68

Number=Defects ≥ 20 weeks or ≥ 500 g CI = Approximate 95% Confidence Intervals

† = RATES BASED ON TOTAL **MALE** BIRTHS

Appendix A.4 Selected Anomalies With Rates Of Live Births (L) and Stillbirths (S) Compared With Total Rates Including Terminations of Pregnancy (ToP), 2000-2005

Table A4.1 2000

Congenital Anomalies (CAs)	Number of Anomalies		Rates/1000 Total Births	
	Live (L) and Still (S)	ToP (T)	L + S	L + S + T
Anencephaly	8	4	0.22	0.33
Spina Bifida	7	4	0.19	0.30
Encephalocele	5	0	0.14	0.14
Hydrocephaly	28	3	0.76	0.84
Rectal & Large Intestinal Atresia/Stenosis	21	5	0.57	0.71
Renal Agenesis/Hypoplasia	19	5	0.52	0.65
Limb Reduction Anomalies	33	6	0.90	1.06
Chromosome Anomalies (all)	106	31	2.88	3.72
Down Syndrome	54	11	1.47	1.77
Syndromes	35	0	0.95	0.95

Table A4.2 2001

Congenital Anomalies (CAs)	Number of Anomalies		Rates/1000 Total Births	
	Live (L) and Still (S)	ToP (T)	L + S	L + S + T
Anencephaly	3	4	0.08	0.19
Spina Bifida	12	1	0.32	0.35
Encephalocele	7	0	0.19	0.19
Hydrocephaly	21	3	0.56	0.64
Rectal & Large Intestinal Atresia/Stenosis	26	4	0.69	0.80
Renal Agenesis/Hypoplasia	19	5	0.51	0.64
Limb Reduction Anomalies	42	10	1.12	1.39
Chromosome Anomalies (all)	108	23	2.88	3.50
Down Syndrome	57	14	1.52	1.89
Syndromes	24	1	0.64	0.67

Table A4.3 2002

Congenital Anomalies (CAs)	Number of Anomalies		Rates/1000 Total Births	
	Live (L) and Still (S)	ToP (T)	L + S	L + S + T
Anencephaly	3	4	0.08	0.18
Spina Bifida	6	2	0.16	0.21
Encephalocele	6	1	0.16	0.18
Hydrocephaly	21	1	0.55	0.57
Rectal & Large Intestinal Atresia/Stenosis	25	4	0.65	0.75
Renal Agenesis/Hypoplasia	25	3	0.65	0.73
Limb Reduction Anomalies	41	3	1.06	1.14
Chromosome Anomalies (all)	111	35	2.88	3.79
Down Syndrome	50	18	1.30	1.77
Syndromes	39	1	1.01	1.04

Table A4.4 2003

Congenital Anomalies (CAs)	Number of Anomalies		Rates/1000 Total Births	
	Live (L) and Still (S)	ToP (T)	L + S	L + S + T
Anencephaly	9	3	0.22	0.30
Spina Bifida	11	3	0.27	0.35
Encephalocele	3	3	0.07	0.15
Hydrocephaly	22	1	0.55	0.57
Rectal & Large Intestinal Atresia/Stenosis	25	2	0.62	0.67
Renal Agenesis/Hypoplasia	18	3	0.45	0.52
Limb Reduction Anomalies	41	10	1.02	1.27
Chromosome Anomalies (all)	114	59	2.84	4.31
Down Syndrome	72	20	1.80	2.29
Syndromes	31	1	0.77	0.80

Table A4.5 2004

Congenital Anomalies (CAs)	Number of Anomalies		Rates/1000 Total Births	
	Live (L) and Still (S)	ToP (T)	L + S	L + S + T
Anencephaly	3	6	0.07	0.22
Spina Bifida	13	2	0.32	0.37
Encephalocele	3	0	0.07	0.07
Hydrocephaly	22	0	0.54	0.54
Rectal & Large Intestinal Atresia/Stenosis	27	7	0.67	0.84
Renal Agenesis/Hypoplasia	22	3	0.54	0.63
Limb Reduction Anomalies	36	9	0.89	1.11
Chromosome Anomalies (all)	129	34	3.18	4.02
Down Syndrome	67	14	1.65	2.00
Syndromes	35	1	0.86	0.89

Table A4.6 2005

Congenital Anomalies (CAs)	Number of Anomalies		Rates/1000 Total Births	
	Live (L) and Still (S)	ToP (T)	L + S	L + S + T
Anencephaly	7	1	0.17	0.19
Spina Bifida	8	4	0.19	0.29
Encephalocele	6	3	0.14	0.22
Hydrocephaly	18	3	0.43	0.50
Rectal & Large Intestinal Atresia/Stenosis	15	3	0.36	0.43
Renal Agenesis/Hypoplasia	15	3	0.36	0.43
Limb Reduction Anomalies	32	13	0.77	1.08
Chromosome Anomalies (all)	154	50	3.68	4.88
Down Syndrome	84	28	2.01	2.68
Syndromes	32	4	0.77	0.86

Appendix A.5 Numbers of Cases, Anomalies and Anomalies per Case 1980-2005 Live Births and Stillbirths

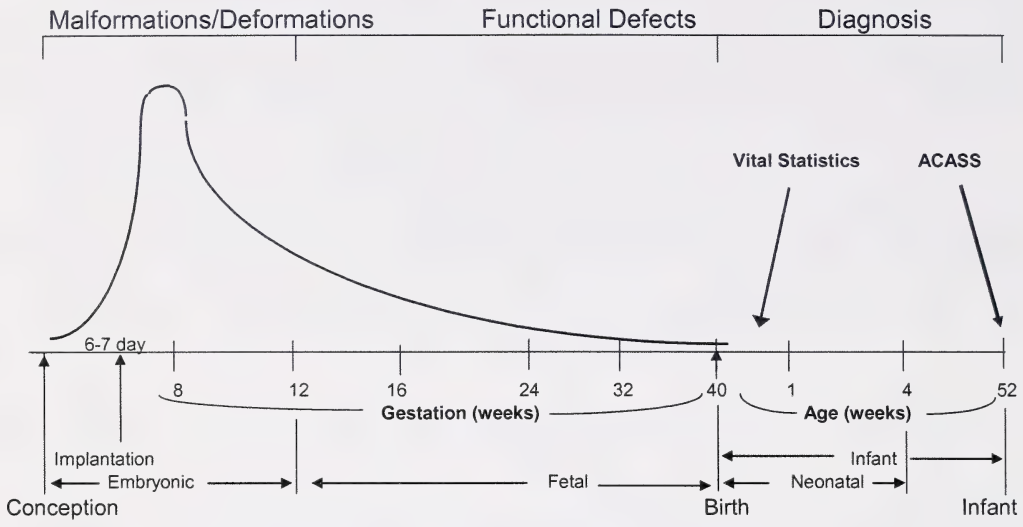
Year	Total births (Live & Still)	# Cases (Live & Still)	Case Rate/1000 Total births	# Anomalies (Live & Still)	Anomaly Rate/1000 Total births	Average # Anomalies/ case
1980	39655	1423	35.88	1782	44.94	1.25
1981	42463	1444	34.01	1903	44.82	1.32
1982	44987	1570	34.90	2133	47.41	1.36
1983	45381	1486	32.74	2133	47.00	1.44
1984	43864	1583	36.09	2167	49.40	1.37
1985	43565	1688	38.75	2439	55.99	1.45
1986	43552	1778	40.82	2440	56.02	1.37
1987	41957	1694	40.37	2432	57.96	1.44
1988	41970	1873	44.63	2767	65.93	1.48
1989	43223	1930	44.65	2891	66.89	1.50
1990	42895	1985	46.28	2972	69.29	1.50
1991	42675	1779	41.69	2577	60.39	1.45
1992	41944	1775	42.32	2647	63.11	1.49
1993	40163	1468	36.55	2200	54.78	1.50
1994	39720	1409	35.47	2165	54.51	1.54
1995	38784	1177	30.35	1873	48.29	1.59
1996	37710	1140	30.23	1777	47.12	1.56
1997	36798	1078	29.30	1832	49.79	1.70
1998	37808	1143	30.23	1889	49.96	1.65
1999	38026	1172	30.82	2200	57.86	1.88
2000	36839	1257	34.12	2123	57.63	1.69
2001	37460	1349	36.01	2314	61.77	1.72
2002	38532	1334	34.62	2255	58.52	1.69
2003	40118	1448	36.09	2433	60.65	1.68
2004	40557	1496	36.89	2682	66.13	1.79
2005	41814	1472	35.20	2507	59.96	1.70
Total	**1062460**	**38951**	**36.66**	**59533**	**56.03**	**1.53**

Appendix A.6 Termination of Pregnancy (ToP) for Congenital Anomalies, 1997-2005

Year of Termination	# ToP cases	# ToP anomalies	Average # Anomalies/ Per Case
1997	65	126	1.94
1998	69	231	3.35
1999	54	183	3.39
2000	53	147	2.77
2001	52	177	3.40
2002	62	136	2.19
2003	91	214	2.35
2004	74	205	2.77
2005	87	210	2.41
Total	**607**	**1629**	**2.68**

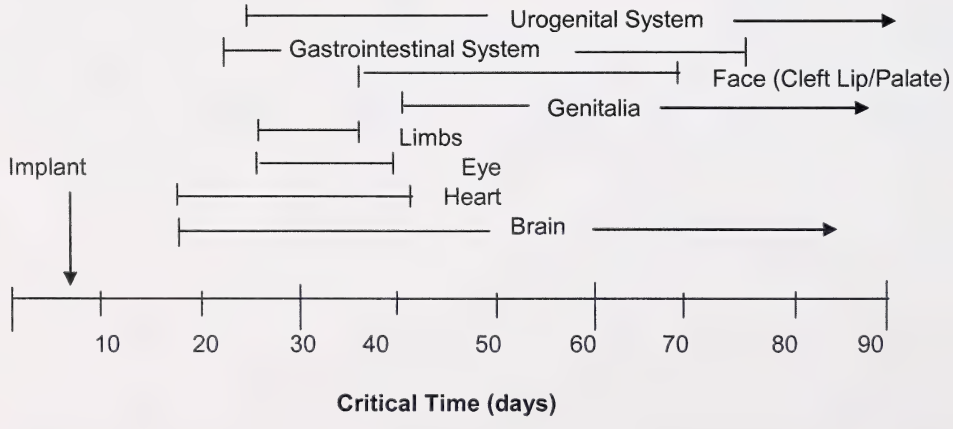
Note: These anomalies are not registered with Vital Statistics i.e. <20 weeks or <500g.

Appendix A.7 Diagram of Embryonic and Fetal Developmental Stage and Diagnosis of Congenital Anomalies



Congenital Anomalies: Embryonic and Fetal Developmental Stage and Diagnosis

Appendix A.8 Critical Periods of Embryogenesis by Major Organs/Systems in Humans



Critical Periods of Embryogenesis by Major Organs/Systems in Human